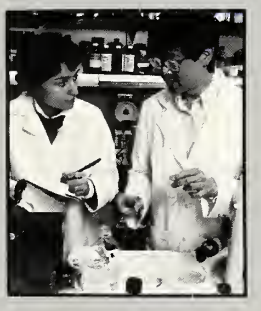
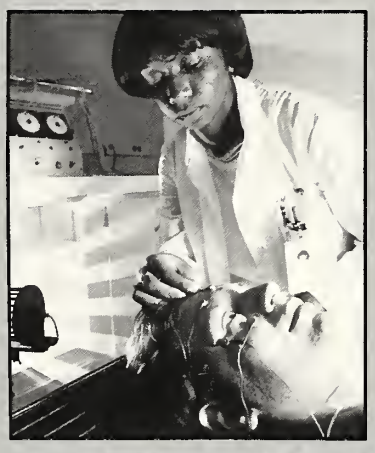
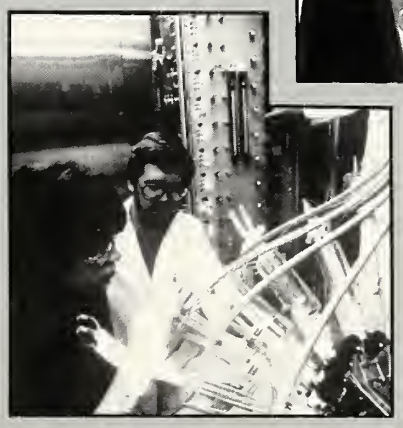
A Report of the
Task Force on the

National Strategic Research Plan







National Institute on
Deafness and Other
Communication
Disorders

April 1989

National Institutes of Health
Bethesda, Maryland

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Task Force on the

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Acknowledgments

The Task Force extends special thanks to Sean Donohue, National Institutes of Health, who coordinated this project; to Tom J. Truss, Jr., Ph.D., the principal science writer and editor of the Task Force report; to Jeffrey Fox, Ph.D., Marlene W. Karakashian, Ph.D., Christy Ludlow, Ph.D., and Cynthia Shewan, Ph.D., who assisted Task Force members in writing numerous draft reports; and to the staff of Social & Scientific Systems, Inc., of Bethesda, MD, which was the support contractor for the entire project.

Foreword

This National Strategic Research Plan is the collaborative result of a Task Force of over 100 highly respected U.S. experts in clinical and basic science from a broad variety of specialties in deafness and other communication disorders, and it is intended to serve many purposes. One is to guide the decisionmaking processes of the National Advisory Board and National Advisory Council of the newly authorized National Institute on Deafness and Other Communication Disorders (NIDCD) as these bodies begin meeting the mandate of Public Law 100-553, which authorized the new Institute. Another purpose is to inform the Nation's scientific investigators of possible areas of opportunity for NIDCD-funded research and to provide them with initial guidance as they plan their own research agendas. The plan will also assist persons with communication disorders and their support organizations in understanding what the Federal Government, through the National Institutes of Health (NIH), has been able to achieve in the past to help them realize their potential and what it might be able to achieve in the future. This document is also a response to P.L. 100-553, which requests that the Director of the NIDCD develop a research program on deafness and other communication disorders and prepare a plan to initiate, expand, intensify, and coordinate the activities of the Institute. In addition, the plan is intended to inform members of Congress of research progress and future research opportunities within the purview of the NIDCD. The document will also assist, in the course of the Institute's day-to-day operations, in providing clarity and direction to the growth of the NIDCD research portfolio.

Members of the Task Force are due special words of thanks for giving unstintingly of their talents and time in seeing this document through to its completion. Their assembly on January 17-19 brought together for the first time representatives of a broad array of scientific disciplines, and they were able to exchange diverse ideas and arrive at new concepts. They worked long, productive hours, often well into the night, in reaching a consensus. During February and March, they refined their efforts with telephone conferences and numerous revised texts.

Drs. Charles I. Berlin and Robert J. Ruben, Task Force cochairs, should be singled out for special gratitude. This National Strategic Research Plan is very much a product of their experience, expertise, and guidance. A decade ago, they were members of the Panel on Communicative Disorders to the National Advisory Neurological and Communicative Disorders and Stroke Council. In developing their 1979 recommendations, they worked with a large number of subcommittee members and consultants, many of whom have also served on this NIDCD Task Force. Now joined with experts and new talent, including those in emerging areas of expansion, they have together provided a link with past achievements and helped formulate this plan for future research.

The Task Force was asked to identify, in its discussions, short-range (3-year) and long-range research goals in deafness and other communication disorders. Where possible, this

division is recognized in the research plan. The rate of research progress, however, often depends on a variety of resources, including the quality of research facilities and the size of the pool and the skills of highly specialized investigators. The Task Force expressed major concerns about facilities and about needs in the current pool of researchers in deafness and other communication disorders and realized that the achievement of many of the goals may have to await the implementation of new or expanded mechanisms, especially for research training. The division of goals into this timeframe of long-range and short-range categories will thus facilitate the NIDCD's future ability to assess research progress and identify further needs and to make programmatic adjustments based on newly acquired knowledge and on changes in the pool of research talent. This document should therefore be viewed as a promising beginning to exciting advances in research on deafness and other communication disorders.

Jay Moskowitz, Ph.D.
Director (Acting)
National Institute on Deafness and
Other Communication Disorders

Preface

The *National Strategic Research Plan of the National Institute on Deafness and Other Communication Disorders (1989)* is a far-reaching scientific response to Public Law 100-553, signed by the President on October 28, 1988. The plan presents the views and research recommendations of over 100 eminent U.S. scientists assembled as a Task Force to address the country's biomedical needs in the communication disorders areas.

The convergence of a number of factors brought about the National Institute on Deafness and Other Communication Disorders (NIDCD). The first is that the nature of our society has changed. We live during times when our economic and social well-being is dependent upon our ability to communicate. As a result, the diseases and disorders within the purview of the NIDCD underlie increasingly widespread societal losses and suffering. Another factor is that the amount of research being conducted in the communications areas, compared to research in other areas, has been small, and was fragmented across a number of Institutes of the National Institutes of Health (NIH). Research on communication disorders was not the primary mission of any of the NIH Institutes. Not all of the current advances in technology were being fully utilized in research and in efforts to apply knowledge to the alleviation and amelioration of the disorders.

In this plan, the Task Force describes the research opportunities where the biomedical sciences can lead to the prevention, cure, and amelioration of many disorders of communication. By combining basic and clinical research with currently available technology, investigators should be able to solve many of the problems of otitis media, nerve deafness, balance disorders, language deficiency, and diseases of smell and taste. What is apparent from the plan is the need to stimulate and expand these areas with an infusion of fiscal and research personnel resources. The most critical need is the recruitment and training of scientists. Their work in laboratories and clinics will bring the current state of scientific knowledge and its techniques to bear on research on these "modern" diseases, which have become quite visible in this, the Age of Communication.

In developing this Strategic Research Plan, scientists in the various areas of communication disorders were brought together for the first time in U.S. history, and they were able to guide and advise each other and the NIDCD on a mutually determined research agenda. Public neglect of these disorders is now coming to an end, and the NIDCD can serve as a single focus within the NIH to implement the research that will expand basic knowledge about them and its application to treatment. For instance, the problems of subcellular interaction of sensory cells, the processes of plasticity of the nervous system in regard to sensory input, and the areas of final effect, such as language, now are all considered in one NIH Institute. This intellectual and programmatic integration of many diverse disciplines will

facilitate more efficient utilization of human and physical resources to study communication disorders.

We wish to thank all those who have worked so diligently and long to bring about the establishment of the NIDCD and those who gave so unsparingly of their time to this Task Force report. It was an exciting and rewarding experience. The National Strategic Research Plan is now complete, and it is the result of the first national scientific consensus on the state of knowledge and research needs and opportunities in the area of communication disorders. In the future, the plan will be reviewed and revised, as new knowledge and new research opportunities emerge. We expect to look back on this publication as a sketch that led to productive coordination and cooperation across disciplines and brought about a betterment of lives everywhere.

Charles I. Berlin, Ph.D.
Robert J. Ruben, M.D.
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Task Force on the National Strategic Research Plan
National Institute on Deafness and Other Communication Disorders

Contents

Foreword iii

Preface v

Executive Summary 3

Chapter 1. Deafness and Hearing Disorders 37

Chapter 2. Balance and the Vestibular System 71

Chapter 3. Voice and Voice Disorders 93

Chapter 4. Speech and Speech Disorders 129

Chapter 5. Language and Language Impairments 157

Chapter 6. Smell, Taste, and Touch 189

Chapter 7. Cross-Cutting Issues 229

Appendix A. Public Law 100-553 255

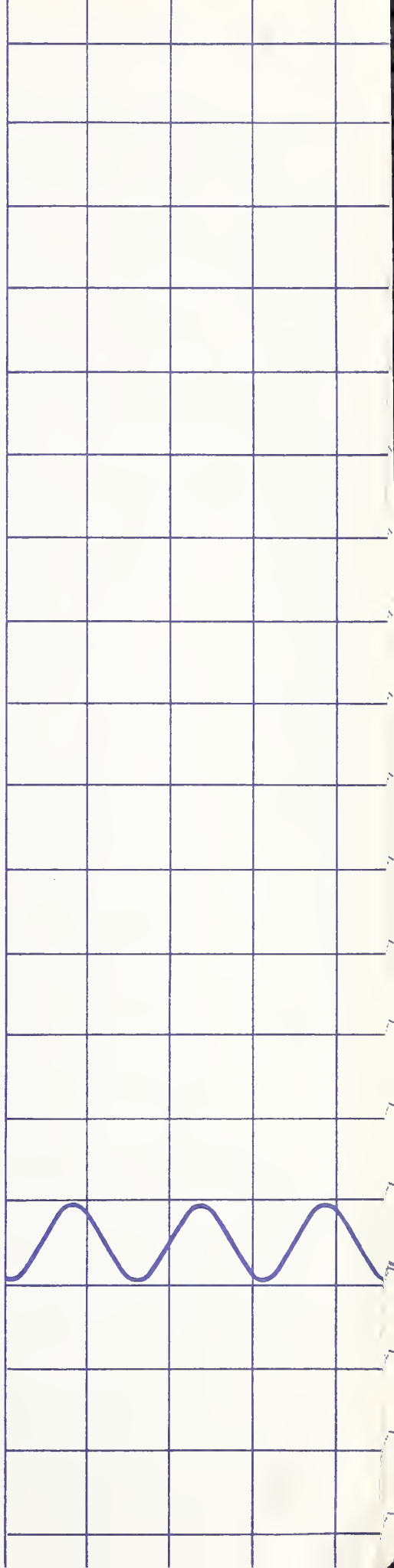
Appendix B. Task Force on the National Strategic Research Plan 265

Appendix C. Reviewing Organizations 289

Appendix D. Comments by Reviewing Organizations 293

Appendix E. A Comprehensive Program of Research on Deafness and Hearing Disorders 333

Executive Summary



CONTENTS

BACKGROUND 7

DEAFNESS AND HEARING DISORDERS 9

Major Scientific Research Opportunities 10

Encoding of Sound by the Inner Ear 10
The Central Nervous System 11

Major Clinical Research Opportunities 11

Genetic Hearing Loss 11
The Bases and Rehabilitation of Severe and Profound Hearing Loss 11
Otology and Rehabilitation 12

BALANCE AND THE VESTIBULAR SYSTEM 12

Major Scientific Research Opportunities 13

Anatomy 13
Physiology 14
Chemistry 14
Psychophysics and Perception 14
Plasticity 14

Major Clinical Research Opportunities 15

Use of Pathology to Elucidate Basic Mechanisms 15
Demographics 15
Diagnostic Methods and Testing Procedures 15
Treatment Efficacy 15

VOICE AND VOICE DISORDERS 16

Major Scientific Research Opportunities 17

Laryngeal and Respiratory Physiology 17
Biomechanics of the Larynx 17
Cellular and Molecular Biology and Anatomy 17
Development, Critical Periods, and Aging 18

Major Clinical Research Opportunities 18

Organic, Idiopathic, and Neurologic Diseases 18
Procedures for Diagnosis and Treatment 19
Instrumentation and Technological Developments for Diagnosis and Treatment . 20

SPEECH AND SPEECH DISORDERS 20

Major Scientific Research Opportunities 21

Normal Anatomy and Physiology 21
Normal Speech Development and Aging 22
Devices for Speech Analysis 22

Major Clinical Research Opportunities 22

Speech of the Hearing Impaired 22
Speech Production in Individuals Who Stutter 22
Speech in Individuals with Craniofacial Anomalies 22
Speech in Neurological Disorders of Communication 22
Research Needs Across Populations 23
Technological Needs Across Populations 23

LANGUAGE AND LANGUAGE IMPAIRMENTS 23

Major Scientific Research Opportunities: Children 24

Normal Processes and Mechanisms 24
Deaf and Language Impaired Children 25
Brain-Behavior Relationships 25

Major Scientific Research Opportunities: Adults 25

Brain-Language Relations 25
Behavioral Investigations of Language 25

Major Clinical Research Opportunities: Children 26

Understanding the Nature of Disorders and Atypical Language 26
Intervention Strategies 26

Major Clinical Research Opportunities: Adults 26

Classification 26
Intervention Strategies and Recovery 27

Major Scientific Research Opportunities: Speech and Sign Perception 27

Speech and Sign Perception in Infants and Children 27
Speech and Sign Perception in Adults 27

Major Clinical Research Opportunities: Speech and Sign Perception 27

Assessment 27

SMELL, TASTE, AND TOUCH 28

Major Scientific Research Opportunities: Smell and Taste 29

Plasticity, Including Development, Regeneration, and Aging 29
Access of Infectious Agents and Toxins to the Brain Through the Olfactory Nerve 30
Receptor Mechanisms and Signal Processing Throughout Central Systems . . . 30

Major Clinical Research Opportunities: Smell and Taste 30

Epidemiology of Smell and Taste Disorders 30
Development of New Diagnostic Tests 31
Affective Dimensions of Taste and Smell 31
Associations Between Chemosensory Disorders and Altered Food Intake in Aging and in Various Chronic Diseases 31
Effects of Environmental Agents on Chemosensory Function 31
Clinical Trials to Evaluate Potential Treatment Protocols and Rehabilitative Strategies 31

Major Scientific Research Opportunities: Touch 31

Tactile Sensory Coding and Central Representation 31
Touch and Its Interaction With Other Senses 31

Major Clinical Research Opportunities: Touch 32

Development of Tactile Stimulators 32
The Haptic Exploration of Objects and Surfaces 32
Tactile Sensory Aids 32
Testing of Tactile Sensitivity in the Mouth 32

CROSS-CUTTING ISSUES 32

Executive Summary

BACKGROUND

The National Deafness and Other Communication Disorders Act of 1988 (Public Law 100-553) establishes within the National Institutes of Health (NIH) the National Institute on Deafness and Other Communication Disorders (NIDCD), and requires that the Director, NIDCD, establish a National Deafness and Other Communication Disorders Program. Included in the program are diseases and disorders of hearing, balance, voice, speech, language, taste, and smell. The Director, NIDCD, is also required to submit to the Director, NIH, a plan for implementing the program. This report and these recommendations of the Task Force on the National Strategic Research Plan of the NIDCD are the response to that mandate.

Public Law 100-553 requires that the program include:

- Investigation into the etiology, pathology, detection, treatment, and prevention of all forms of disorders of hearing and other communication processes through the support of basic research in such areas as anatomy, audiology, biochemistry, bioengineering, epidemiology, genetics, immunology, microbiology, molecular biology, the neurosciences, otolaryngology, psychology, pharmacology, physiology, and speech and language pathology.
- Research into the evaluation of techniques and devices used in diagnosis, treatment, rehabilitation, and prevention of these disorders.
- Research into prevention and early detection and diagnosis of these disorders, including disorders in the elderly.
- Research to expand knowledge of the effects of environmental agents that influence hearing and other communication processes.
- Development and facilitation of intramural programs of research on clinical and fundamental aspects of disorders of hearing and other communication processes.

P.L. 100-553 also requires the establishment of a data system for the collection, storage, analysis, retrieval, and dissemination of information derived from patient populations, a national information clearinghouse, and multipurpose research centers. P.L. 100-553 can be found in its entirety in Appendix A.

The enactment of P.L. 100-553 is especially timely. In the past, the disorders covered by the act were recognized only in their most extreme forms, such as profound deafness or the entire absence of speech. Many of the disorders have been defined

only recently as separate entities. A few decades ago, a child with a language impairment was often considered mentally retarded, and a person who stuttered was believed to have a psychological problem. Other disorders were not recognized as conditions requiring medical or rehabilitative intervention. For instance, a child with a moderate hearing loss, an elderly person with an unsteady balance, or a teacher with a hoarse voice was dismissed as not being impaired sufficiently to need professional attention.

This set of attitudes has changed. As previously reported to Congress, a large number of infants and small children have disorders and diseases affecting their hearing and speech. Of every 22 infants born in the United States, one has, or shortly will have, a hearing problem. Ear infections are the most prevalent disease of young children from birth to age 7. At least 1 million children are deaf or have other communication disorders. Such children are often left living in a world apart from others throughout the formative years of their lives. They are often unable to read beyond the level of grade 6 because their ability to learn language has been compromised.

Communication disorders also exact a large economic toll. Disorders of hearing, speech, and language alone cost the U.S. economy an estimated $30 billion a year in lost productivity, special education costs, and medical costs.

To help the NIDCD develop a National Strategic Research Plan, a Task Force of experts, representing scientific disciplines that span the biomedical and behavioral research spectrum from basic research to clinical practice, was convened on January 17-19, 1989. Members of the Task Force are listed in Appendix B. They were asked to develop a report and recommendations for research in the topic areas represented in chapters 1 through 6 of this publication: deafness and hearing disorders; balance and the vestibular system; voice and voice disorders; speech and speech disorders; language and language impairments; and smell, taste, and touch. In each of these areas, the Task Force reviewed the following:

- Historical overview of the field, including prevalence, incidence, and cost;
- Recent research accomplishments;
- Program goals; and
- Research opportunities, strategies, and priorities, formulated on short-range (3 years) and long-range bases.

This time frame was established to provide an opportunity for a review of the accomplishments of the program after three years and to make adjustments where they are needed.

The Task Force also reviewed mechanisms for implementing the NIDCD research program, and its recommendations are presented in chapter 7. These recommendations cut across many of the issues raised in the scientific discussion in chapters 1 through 6 and focus on trans-NIDCD concerns.

Following the Task Force's completion of its final draft, the Report was reviewed by agencies and organizations concerned with deafness and other communication

disorders, for suggestions and comments. This action was taken to ensure the broadest possible base and the representation of all points of view in the final Report. The reviewing agencies and organizations are listed in Appendix C. Their responses are reprinted in Appendix D.

DEAFNESS AND HEARING DISORDERS

Chapter 1 focuses on deafness and hearing disorders, which affect an estimated 28 million persons in the United States, including approximately 2 million persons who are profoundly deaf. The most common cause of hearing loss is otitis media, which is primarily a disease of infants and young children. The disease accounted for 10 million visits to doctors' offices in 1975 and costs the economy an estimated $1 billion per year. Chapter 1 observes that hearing impairments are invisible and nonfatal and, as a result, the disorders attracted little public attention in the past, and research on hearing and hearing impairments lagged behind research in other areas. In addition, there has been a long history of societal ignorance about deafness. This attitude has changed in recent years, and remarkable progress has been made in understanding hearing disorders. It has been learned, for instance, that the cochlea, which is located in the inner ear and is the seat of the organ of hearing, is not a passive converter of acoustic energy, or sound. It also *produces* acoustic signals, which participate with ear hair cells in a feedback mechanism, that enhances the "hearing" action of the inner ear. Scientists have named these internally produced signals *otacoustic emissions*. Disruption of this feedback leads to hearing loss. Also, much has been learned, through labeling techniques and electron microscopic examination, about the nerve fiber populations that connect the ear with the brain.

Other areas in which advances have been made include the understanding of how the perception of sound is localized in space. Such findings have come largely from studies of bats and barn owls. Nonprimate studies have also led to the unexpected finding that sensory cells in cold-blooded animals regenerate after birth and can be regenerated even in species where their production normally ceases in early embryonic development. Results of experiments using recently developed methods of biotechnology may lead to a revolutionary change in the widely held view that human "nerve" deafness is irreversible. Much has also been learned about otitis media, especially with regard to immune mechanisms and their genetic regulation, and about the protective roles of lysozyme and lactoferrin, which are naturally occurring substances in the mucous system of the eustachian tube.

Major advances have been made in noninvasive diagnostic procedures to detect disorders of the ear. The technique called immittance audiometry records signals produced by middle ear muscles, and use of the technique has increased the likelihood of identifying the site of lesions in the middle ear. Another new technique, the

auditory brain stem response (ABR) test, has improved the ability of investigators to assess the integrity of the auditory pathways of the brain. Such tests provide information about the response of certain brain portions of the auditory system to experimentally presented stimuli. The technique is especially useful in detecting hearing loss in neonates. Ongoing investigations are determining whether magnetic resonance imaging (MRI) and positron emission tomography (PET) can detect major abnormalities in the function of auditory structures in the central nervous system.

In the area of basic studies, much has been learned about the function of the cochlea, such as measuring cochlear blood flow and describing its regulation. There is now a greater understanding of the biophysics of mechanoelectrical transduction in hair cell membranes. In addition, cells and tissues from the inner ear can now be grown in culture and are available for in vitro studies. Membrane properties of auditory nerve fibers can be analyzed by manipulating their electrical and chemical environments. New techniques are demonstrating the importance of head and torso position for the localization of sound and are providing new possibilities for presenting sounds, such as through hearing aids.

In the area of environmentally induced hearing loss, important basic information is now available about the effects of noise on the microcirculation of the cochlea and the mechanical properties of sensory cells.

In the area of clinical studies, genetic analysis of some deaf families is under way. Genetic markers have been identified that are predictive of risk of otitis media. Also on the horizon is the possible application of monoclonal antibody techniques to assist in diagnosis by discriminating between normal and abnormal cell populations in the auditory system.

In the area of prosthesis development, controversy about several aspects of cochlear implants continues, including the determination of appropriate candidate patients, device selection, preimplantation and postimplantation assessments, and rehabilitation procedures.

Major Scientific Research Opportunities

Encoding of Sound by the Inner Ear

- Analyze receptor cell activity in simple and complex sounds (including speech) and how this activity is modified through brain-controlled feedback systems.
- Elucidate the cellular and molecular mechanisms that underlie active processes that determine the extraordinary sensitivity and exquisite selectivity of the inner ear and the basis for otacoustic emissions—that is, sounds that originate from the ear.
- Define the relationship between information in the sound signal and coded activity of the auditory nerve.
- Determine the homeostatic and immunological mechanisms (including blood flow, metabolism, and

biochemical features) that maintain the normal environment of the inner ear and factors that may control regeneration and repair of sensory elements of the inner ear.

The Central Nervous System

- Define the representation and processing of signals (including speech) in the central auditory pathways, the structural and synaptic mechanisms that underlie this processing, and how these activities result in perception.
- Assess the normal life cycle of the auditory system, including the definition of the range and critical periods involved with development of auditory processes and the limits of neural plasticity in the auditory system, and define the influence of environmental and nutritional factors as well as pathological factors that compromise and distort the normal life cycle of the system.
- Encourage and support the recruitment of information and techniques from other disciplines—for instance, signal processing, artificial intelligence, molecular and cellular biology, and imaging technology.

Major Clinical Research Opportunities

Genetic Hearing Loss

- Localize and characterize the genes responsible for deafness.
- Conduct populations studies to identify the epidemiological factors of genetic deafness.

The Bases and Rehabilitation of Severe and Profound Hearing Loss

- Conduct investigations in the area of sensorineural hearing loss, in which etiological factors, the study of distortions in perceptual space, as well as the potential for sensory cell regeneration are determined.
- Continue development and assessment of the cochlear prostheses, as well as other sensorineural prostheses of the auditory system, clearly showing promise for providing speech discrimination for a significant percentage of individuals who now suffer complete isolation from the world of sound. These efforts should include studies of enhanced signal processing techniques for the development of neural prostheses as well as hearing aids that can benefit a very large percentage of the hearing impaired.
- Determine life cycle risk factors of sensorineural hearing loss, and conduct clinical trials to define the efficacy of proposed therapeutic procedures to reduce or reverse it.
- Study middle ear disease processes, including those of the inner ear bone, and the cellular and molecular mechanisms involved with otosclerosis. Also develop biochemical and immunological

treatments that may inhibit this disease process, with clinical trials to assess various treatments of middle ear disease. In addition, develop vaccines to eliminate middle ear infections.

- Develop a better understanding of the basis for tinnitus and management strategies to alleviate this disorder.

Otology and Rehabilitation

- Conduct technical and behavioral studies to enhance the development of hearing aids as well as other assistive aids and their fitting for the hearing impaired.
- Define and assess the consequences of hearing disorders and the changes in perception with inner ear hearing loss, particularly in regard to the hard-of-hearing child, and the manner in which these changes may affect normal speech and language acquisition and educational development.

BALANCE AND THE VESTIBULAR SYSTEM

The subject of chapter 2 is balance and the vestibular system. Over 90 million Americans, age 17 and older, have experienced a dizziness or balance problem. In the United States, there are an estimated 97,000 cases of Meniere's disease each year, which affects the inner ear and causes episodes of vertigo, fluctuating hearing loss, and tinnitus (ringing in the ear). The approximate physician charges for visits for these disorders in 1976 totaled $500 million.

The primary receptors of the vestibular system are located in the inner ear, and the system itself is responsible for the sense of orientation in space, for maintaining balance and posture, and for the ability of a person to keep an object in focus when the object or the person is in motion. The vestibular detectors and their connections in the central nervous system allow the brain to integrate information from the eyes, skin, muscles, and joints to create what is perceived to be body orientation, and being at rest or in motion. Because the system has many components, there are many different symptoms that reflect its malfunction.

A current view of the vestibular system is that the vestibular receptor organs "process" the forces associated with head acceleration and changes in head position relative to gravity. Nerves going to brain control centers are excited, and the brain uses these signals to develop a sense of orientation and to activate the muscles that subserve automatic movement of the eyes, locomotion, and posture. Many of the vestibular pathways in the central nervous system are organized into reflex pathways that serve to stabilize and coordinate movements of the eyes, head, and body. These processes are collectively referred to as the vestibular reflex systems. One such reflex, the vestibular-ocular reflex, causes the eyes to move in reaction to head motion.

Orientation is a complex function that allows a person to know the relationship of

parts of the body to the environment in a dynamic and reciprocal interaction. Maintaining orientation and balance requires the integration of information from many sources, especially the ears, eyes, skin, muscles, and joints, and transforming these signals into coordinated patterns of muscle activity. Because vestibular signals interact with all the major sensory systems and involve major brain centers, a large number of disease processes can impair balance.

Motion sickness can occur with exposure to conflicting orientation cues, perceptions of space, and motion signals. A high incidence of motion sickness occurs in pilots being trained in stationary flight simulators. Trainees learn behavioral responses to prevent the development of motion sickness. Space motion sickness, which affects one-half of astronauts, is a major unsolved problem for space travel.

During the past decade, new protocols and computer-based techniques have been developed that can help in the evaluation of gaze and balance disorders. Precise, quantitative measurements of vestibular reflexes, posture, and the interactions of different sensory inputs to the central nervous system can be measured. Perhaps the most remarkable property of the vestibular and balance control systems is their ability to maintain useful functioning responses to many novel motion environments, and their ability to adapt to abnormal function in one or more of their components. Recent accomplishments include a recognition of the essential role of adaptive plasticity in the normal function of the vestibular system, development of improved mathematical models of vestibular reflexes, and an anatomical description of the basic circuitry in the brain stem subserving the vestibular-ocular reflexes.

Advances in diagnosis include newly available computer-based technology that allows identification, quantification, and localization of the source of vestibular and balance disorders, magnetic resonance imaging to detect space-occupying lesions and several degenerative brain disorders, and dynamic computerized tomography to permit the evaluation of blood flow in various parts of the brain stem and brain. Such advances in diagnosis have led to advances in treatment. Smaller acoustic neuromas and other tumors, for example, can now often be discovered and removed while hearing is still serviceable. Some autoimmune disorders and ataxia syndromes can now be diagnosed and treated with appropriate medication.

Major Scientific Research Opportunities

Anatomy

- Elucidate the microscopic anatomy of receptors (structure, innervation patterns, molecular biology).
- Correlate the morphological characteristics with the neurophysiological characteristics of neurons within different parts of the vestibular nuclei during isolated and combined canal, otolith, and visual stimulation.

- Define the distribution and convergence patterns of different vestibular receptors onto the vestibular nuclei neurons, both anatomically and functionally.
- Define the convergence patterns of pathways conveying information from different sensory systems (visual, proprioceptive) and other brain structures (reticular formation, cerebellum, and higher motor and sensory centers) onto the vestibular nuclei neurons.

Physiology

- Elucidate the biophysical properties of the hair cells and specializations within different regions of the canal and otolith end organs.
- Develop an understanding of the types of physical stimuli that govern adaptive behavior, such as the relative importance of different inputs (for example, inputs from the foveal versus retinal periphery, inputs from visual system versus vestibular end organs, and inputs created by active versus passive motion).
- Develop experimental models for peripheral vestibular dysfunction, and take advantage of species differences and genetic variations.

Chemistry

- Characterize the biochemical properties of vestibular hair cells, including transmitter chemistry.
- Define the action of pharmacologic agents on end organ structure.
- Determine the transmitters and modulators for the vestibular neurons, projections to the vestibular nuclei, and projections to and among higher central nervous system nuclei subserving balance and orientation.

Psychophysics and Perception

- Investigate psychophysical aspects of normal vestibular function, and develop standards for evaluation of normal vestibular sensations.
- Conduct psychophysical studies to understand better the imprinting of vestibular experiences in the development of psychological profiles in normal subjects and patients.
- Evaluate the patient-reported relationship between vestibular symptoms, objective balance abnormalities, and panic disorders.

Plasticity

- Define the anatomical, physiological, and biochemical changes in the neural circuitry that underlie adaptive plasticity.
- Define the basic mechanisms of compensation for injury to different components of the balance system.
- Investigate balance problems in special environments, such as under water, in high-performance aircraft, and in space.

Major Clinical Research Opportunities

Use of Pathology to Elucidate Basic Mechanisms

- Further develop genetic models in animals of human vestibular dysfunction.
- Investigate mutants with auditory anomalies to determine possible associated vestibular anomalies.
- Develop genetic mapping techniques for congenital and neoplastic balance system disorders, using recently developed molecular biology techniques to study the pathology and pathophysiology of balance disorders, with special emphasis on disorders of unknown etiology.

Demographics

- Establish normative histological and immunocytochemical data on temporal bones and central vestibular nervous system tissues in the same individuals.
- Establish a quantitative profile of normal vestibular reflexes, including postural development in children.
- Establish data banks to assess the distribution of vestibular disorders among different sectors of the population.
- Identify toxic agents in the occupational environment and in therapeutic agents that impair the balance system.

Diagnostic Methods and Testing Procedures

- Determine the sensitivity, specificity, and clinical efficiency of all present and future diagnostic methods for all major categories and classifications of vestibular abnormalities.
- Encourage and facilitate the development of new diagnostic methods and procedures for documentation of vestibular and other balance disorders.
- Develop diagnostic procedures based on measurements of interrelated auditory and vestibular systems, such as electrocochleography and middle ear biophysical measures.
- Develop a model for computer simulation of sensations associated with balance disorders in order to establish objective measures of symptoms.
- Develop functional tests of otolith organ, vertical semicircular canal interactions, and interactions of these sensory inputs in the central nervous system.

Treatment Efficacy

- Evaluate pharmacologic therapies to insure that they are safe and effective in a variety of settings, including hazardous work environments.
- Establish outcome analysis methods for the medical and surgical management of major disorders of the vestibular and balance systems.

- Evaluate balance training and movement strategy exercises for possible use in at-risk populations.
- Develop and evaluate assistive devices for balance disorders.

VOICE AND VOICE DISORDERS

Chapter 3 provides a discussion of voice and voice disorders. Voice science is a relatively new and growing field, and there are no reliable data on voice disorders among postpubescent populations. Within the last decade, however, the field and the clinical specialties of phonosurgery and voice therapy expanded in a variety of ways. A large new knowledge base and clinical delivery system have developed, resulting in recognition, diagnosis, and treatment of patients with voice impairment and laryngeal pathology. The larynx, or voice box, keeps vocal folds (the vocal chords) open during breathing so that air can enter the lungs. During swallowing, the vocal folds close tightly. The cough is a protective reflex in which vocal folds close tightly and then open abruptly. This action prevents food or liquid from entering the lungs. Voice, or phonation, is generated when airflow from the lungs is modulated by vibrations of vocal folds in the larynx. When vocal fold vibration is impaired, sound generation for speech is affected. Laryngeal movement disorders include spasmodic dysphonia (abnormal movement of muscles in the vocal folds), vocal tremor (rhythmic contractions of laryngeal muscles—a "shaky voice"), and laryngospasm (uncontrolled closing of the vocal folds).

New technologies for voice science have been developed. In videofiberoptic nasopharnygoscopy, video recording of the larynx and vocal folds can be made during speech, swallowing, and breathing. In laryngeal stroboscopy, paced bursts of light synchronously timed with vocal fold vibrations provide a slow-motion view of vibrations of the folds during speech. In addition, a number of new noninvasive tools to perform acoustic measurements provide parameters that reflect voice disorders. New measurement tools are also being developed to provide physiologic and airflow information, which will lead to theoretical models of vocal focal vibration that include airflow dynamics, tissue strain, and vocal fold structure. Magnetic resonance imaging is now being used to measure changes in the shape of the vocal tract. The information should improve the understanding of how phonation becomes speech. In addition, laryngeal muscle activations are now being studied to determine the biomechanics of laryngeal muscle forces during breathing, airway closure, swallowing, coughing, singing, and speaking.

The newly developed acoustic measures of voice can now be used to screen persons for early signs of laryngeal cancer, and new surgical techniques (phonosurgery) are being used to improve and restore voice by removing benign growths, correcting structural abnormalities, and repairing

trauma. Spasmodic dysphonia—once thought to be a psychiatric disorder—can now be diagnosed with videolaryngoscopy, and experimental treatment using botulinum toxin has recently been found to be effective.

Major Scientific Research Opportunities

Laryngeal and Respiratory Physiology

- Evaluate the compartmentalization of the laryngeal muscles with regard to histochemistry and motor unit organization during different gestures, such as inhalation, exhalation, effort closure, phonation, and swallowing.
- Obtain information about the modification of sensory and muscle action feedback to the motoneuron pools during different laryngeal gestures in higher level primates and humans.
- Map the cortical control of the motor and sensory aspects of the larynx.
- Study the regulation of pitch, loudness, register, and quality, with emphasis on the role and interactions of specific muscles and on their aerodynamic properties in relation to changes in the length, shape, configuration, and forces of the vibrating structures. Include vocally trained subjects (singers, actors, public speakers) in addition to normal and impaired subjects.
- Identify the mechanisms of vocal fatigue, and develop theories about the processes of recovery.
- Undertake studies that compare human laryngeal neurophysiology with that of other species proposed as models for human voice production, and explore the validity of study of laryngeal neurophysiology in higher level primates.

Biomechanics of the Larynx

- Investigate the mechanical properties of skin, cartilage, fat, ligament, tendon, and muscle tissues of the larynx.
- Study acoustic and biomechanical source-vocal tract interactions, particularly the effects of vertical movements of the larynx and source filter effects.
- Combine recent developments in the theory of chaos (a new field of study that attempts to find order in seemingly random behaviors) with more traditional linear and nonlinear analysis to describe vocal fold vibration.

Cellular and Molecular Biology and Anatomy

- Acquire new information, which can be developed with existing cellular and molecular biology techniques of immunocytochemistry and in situ hybridization, about the distribution of extracellular matrix molecules, oncogenes, growth factors (for example, epithelial growth factor), and growth-factor

receptors, as they relate to normal structure of the larynx.

- Conduct embryology studies of epithelial-mesenchymal tissue interactions that are the driving force behind normal development of the larynx, with attention to how these interactions affect cell differentiation and final organotypic form.

Development, Critical Periods, and Aging

- Characterize voice production as it relates to the developing structure, with emphasis on the regulation of pitch, loudness, register, and vocal quality, with changes in morphological structure.
- Identify the critical periods for optimum development of vocal control, including infancy, puberty, time of auditory impairment, and periods associated with hormonal change, as well as periods when a speaker is recognized as being at risk for developing dysphonias.
- Study the use of physiological feedback and motor learning in training vocal control in the congenitally deaf and in adults who are profoundly deaf.
- Establish the nature of geriatric voice changes, and develop techniques to forestall or prevent them when such changes may interfere with a person's life or livelihood.
- Determine how selective cell death and tissue atrophy affect respiration and voice in aging.
- Collect anatomical, cellular, and molecular information from aged larynxes of a genetically defined species, with emphasis on localization of extracellular matrix components, growth factors, oncogenes, and growth-factor receptors.
- Expand the base of information about the cell biology of joint deterioration in the aging larynx.
- Determine how to improve the secretory process leading to increased pulmonary efficacy.

Major Clinical Research Opportunities

Organic, Idiopathic, and Neurologic Diseases

- Improve the understanding of inflammatory diseases of the upper airway and larynx, including rhinitis, sinusitis, and laryngitis, and develop better therapy.
- Undertake viral typing of laryngeal papillomas and assess their recurrence rate, effects of treatment, effects of cofactors such as smoking and voice abuse, and rates of association with carcinoma of the larynx.
- Evaluate long-term treatment of spasmodic dysphonia and vocal fold tremors with minute injections of botulinum toxin into the intrinsic laryngeal muscles to determine whether this treatment continues to be effective with repeated injections over several years.

- Study the neural control of the larynx to determine why laryngeal functions are particularly vulnerable to neurological disorders causing selective neuronal loss either in the brain stem or in the basal ganglia.
- Clarify the relationships between neuropathology and laryngeal disorders.
- Expand the knowledge about the mechanisms underlying the formation of idiopathic dysphonias, including environmental noise, air pollution, and systemic disease.
- Conduct epidemiological surveys and clarify the precise etiology and pathophysiology, and strategies for prevention, diagnosis, and management of the effects of environmental agents on the upper respiratory tract (nose and sinuses) and the larynx.
- Expand the information about the effects of reduced respiratory capacity on speech and on the neuromuscular mechanisms and reflexes regulating respiratory volume and flow during speech.

Procedures for Diagnosis and Treatment

- Expand basic science and clinical information about the pathophysiology, prevention, assessment, and treatment of problems associated with laryngeal surgery and injury.
- Clarify the best strategies for evaluating and treating patients with vocal disorders due to the effect of hormonal and pharmacologic substances on the voice, and find new strategies for prevention.
- Continue systematic investigation of voice production using kinematic, acoustic, aerodynamic, and imaging diagnostic aids to determine which technique (or combination of techniques) is most useful in detecting disorders, documenting changes resulting from treatment, identifying persons at risk for developing voice disorders, setting standards for routine assessment, and improving the understanding of the process of voice production.
- Establish reliable methods for objective voice measurement, including measures of vibratory function, acoustic measures, phonatory function analysis, airflow, improved electroglottography, and repeatable psychoacoustic measures, and identify appropriate tests and develop a convenient, inexpensive equipment armamentarium for performing them so that they may be used by practicing otolaryngologists and speech-language pathologists, and, to some degree, in conservatories and acting schools.
- Obtain further electromyogram data, during different vocal control tasks, on various neurogenic-based dysphonias, and in conjunction with other techniques.
- Conduct human and animal studies of selective intrinsic laryngeal muscle denervation to gain improved understanding of the potential role of laryngeal impairment in many cases of dysphagia.
- Continue development of electronic artificial larynges (external, internal, and

implantable) and tracheoesophageal shunts (with and without alloplastic voice prostheses).

- Expand the capability to electrically pace the paralyzed vocal fold to reestablish its breathing function.
- Expand the use of phonosurgical methods through standardized measurements of vocal output before and after surgery, and of simulation modeling to predict vocal outcome before surgical manipulation.
- Establish, through interdisciplinary teams including teachers of singers, the efficacy of various therapeutic techniques for professional and nonprofessional voice users.

Instrumentation and Technological Developments for Diagnosis and Treatment

- Conduct studies of major categories of voice disorders to determine if sufficiently distinctive profiles make it possible to use low-cost acoustic analysis devices for detection and characterization of specific disorders.
- Determine how measures of air pressure and airflow categorize different pathologies, vocal fatigue, test tasks, changes in vocal control strategies, glottal configurations, and other aspects of vocal function.
- Relate observations made from a combination of imaging techniques to each other through appropriate models for use in understanding voice production and application to clinical populations.

SPEECH AND SPEECH DISORDERS

The immensity of the motor learning task that infants must achieve in learning to speak is evidenced by the high frequency of speech production disorders in young children. Speech and speech disorders are the subject of chapter 4.

On entry to first grade, 5 percent of the child population have speech disorders, such as omitted sounds, substituted sounds, and stuttering. The vast majority of these disorders are idiopathic. Estimates of the prevalence of stuttering among school-aged children range between 0.5 percent and 2.2 percent of the total population. About 40 percent of clients seen by school speech clinicians have disorders of articulation, and 99 percent of clinicians see such children. Although legislation requires States to provide intervention to assist children to develop speech normally, about 20 percent remain speech impaired for the rest of their lives. Even the slightest hearing impairment can interfere with speech learning, and about 0.1 percent of children have hearing impairments that significantly interfere with the development of normal speech. Speech development is also disturbed by cognitive disorders such as mental retardation.

Studies of normal adult speakers have demonstrated the complexities of the speech motor control system. Technological advances have provided instrumentation for measuring lip, jaw, velar, tongue, pharyngeal, and laryngeal movements during speech production. Investigators can now record activity of many of the muscles used to produce these movements. Studies show that speakers frequently use very different combinations of muscles to produce the same speech sound, and that all speakers use slightly different movements to produce the sounds that listeners identify as the same sound. These phenomena are known as motor and perceptual equivalence and are evidence for the extensive computational complexity of the human speech perception and production systems.

Once the highly sophisticated skills of speech have been learned, persons no longer rely on hearing their own or others' speech to produce normal speech. If a normal speaker produces an error in one articulator, other articulators automatically change their movements to compensate. Once speech is learned, it is highly resistant to disturbances due to loss of hearing or changes in sensation. In contrast, changes in the motor control system can have devastating consequences. Different regions of the central nervous system control different aspects of speech production.

Significant advances have been made in the study of stutterers. The fluent speech of stutterers differs from that of normally fluent speakers in many subtle ways. Neurophysiological differences in brain function may interfere with speech production in adult stutterers. In addition, genetic studies suggest a genetic predisposition for stuttering in some families.

Talking machines, using microcomputers, have recently become available for persons with severe speech impairments—those "locked in" by cerebral palsy, laryngectomy, neurological disease, and other disorders. Dozens of new devices help patients converse, use the telephone, and respond in the classroom.

Recently developed microcomputers can automatically analyze speech in real time and produce a visual display on a screen while a speaker is saying words and sentences. These systems are now being used to train deaf children to speak.

Techniques such as miniature magnetometers and ultrasound are being used to study tongue, pharyngeal, and velar movements during speech, chewing, and swallowing. With computerized signal processing, analysis of movements, muscle action, and speech waveforms is now feasible in both the laboratory and the clinic.

Major Scientific Research Opportunities

Normal Anatomy and Physiology

- Conduct population-based studies of gross structure of the vocal tract.
- Characterize tissue properties of muscles of articulation.
- Study peripheral and central neural mechanisms of the speech system.

Normal Speech Development and Aging

- Conduct large-scale retrospective and prospective studies of speech development in normal children, comparing normal populations with those at risk for communicative disorders.
- Study the effects of normal aging on speech development, so that abnormal signs can be readily detected at any time in the life cycle.

Devices for Speech Analysis

- Develop and apply new techniques for static and dynamic imaging of the vocal tract, including magnetic resonance imaging, x-ray microbeam, magnetometer systems, ultrasound, and dynamic palatography.
- Develop articulatory synthesis techniques.

Major Clinical Research Opportunities

Speech of the Hearing Impaired

- Conduct large-scale studies of the effects of hearing loss at different stages of the life cycle on speech production.
- Investigate factors with major effects on the intelligibility of the speech of the hearing impaired as a function of educational history and residual hearing.
- Develop speech training aids, with protocols for their use. Such aids must deliver articulatory information in a focused manner and assess learning independently for different components of the articulatory system.

Speech Production in Individuals Who Stutter

- Conduct studies focused on discoordination among respiratory, laryngeal, and articulatory systems.
- Investigate the effects of standard therapeutic regimens on various aspects of articulation.
- Investigate stuttering symptom constellations, with an objective of delineating subgroups of individuals.

Speech in Individuals with Craniofacial Anomalies

- Study the clinical history of patients with orofacial anomalies, with careful grouping by genetic syndrome. The full range of communicative defects should be assessed before and after treatment.

Speech in Neurological Disorders of Communication

- Develop standardized assessment protocols, based on perceptual, acoustic, and physiological data, for neurological disorders of communication.

Research Needs Across Populations

- Develop basic epidemiological information on the prevalence and risk of speech disorders in multicultural populations, with emphasis on nondiscriminatory assessment and effective service delivery.
- Conduct long-term studies of school-age populations assessing relations among speech and language disorders.

Technological Needs Across Populations

- Develop a data base of microcomputers in use in research and service delivery.
- Conduct "real life" studies of characteristics of client populations for augmentative communication, their interaction with existing devices, and unmet needs. Results of such studies should guide device development.

LANGUAGE AND LANGUAGE IMPAIRMENTS

Language impairment is a disorder of comprehension or production of spoken language sufficient to impair interpersonal communication. It is estimated that between 6 and 8 million people in the United States have some form of language impairment. Chapter 5 discusses language and language impairments across the life cycle, including language acquisition and use by deaf and hearing impaired populations. Three categories of children are discussed: those with specific language impairment, those with acquired language impairment, and those with autism. Language disorders in adults are also reviewed, including disorders associated with Alzheimer's disease and other dementias.

Early studies of the neurological status of specifically language impaired (SLI) children found evidence for frank neurological impairment in only a minority. Neuropsychological studies demonstrated that SLI children performed especially poorly on perceptual and memory tasks. Research in recent years has begun to address problems of inadequate descriptions of subjects. Projects devoted to distinguishing among subgroups of SLI children are now under way. New technologies are beginning to be applied to SLI children to determine whether previously unidentified neurological disorders might be found. Genetic studies have also been undertaken. Evidence is accumulating that young SLI children are at risk for later deficits in reading and that SLI and reading disabled children represent overlapping populations.

Significant advances have been made in the study of children with acquired brain injury. Investigations into the relationship between lesion location and language ability have begun.

Although communication deficits remain a hallmark of autism, it appears that

certain nonlinguistic deficits make a separate contribution to the impaired interpersonal skills of autistic children. Studies of the language of autistic children have focused on comprehension strategies, on spontaneous use of syntax, and on the utility of augmentative and alterative communication techniques.

The clinical populations of language disorders in adults span a wide range of etiologies, from head injury to Alzheimer's disease. Research over the last decade has been particularly productive for a number of brain-related language disorders. Fine-grained analysis of computerized tomography and magnetic resonance imaging scans has permitted the differentiation of previously unrecognized subtypes of aphasia, the identification of lesions associated with specific deficits, and some predictors of recovery or of lasting impairment. High points in remediation of language disorders include the demonstration that profoundly aphasic patients can learn a computerized technique for exchanging information. In addition, results of pharmacological treatment show promise in ameliorating some forms of aphasia.

Ongoing study of speech and sign perception will lead to knowledge about the biology of language, the interaction of biology and the environment in the acquisition of this complex system of symbols, and how speech and sign are ultimately organized in the brain. Comparisons of the processes of speech perception and sign perception in normal and language-disorder hearing and deaf individuals will provide a unique means of determining those aspects of language that are independent of mode (signed or spoken) of communication.

During the past decade, the understanding of normal infants' processing of speech has advanced greatly. It is now known that normal infants' abilities to hear differences in speech are very good at birth, and at 6 months, infants can discriminate sound units not present in their own language. Also at 6 months, infants can recognize a particular speech unit when it is spoken by different talkers using different tones of voice. By 20 weeks of age, infants show a primitive ability to lipread—they detect a match between the presentation of a sound and the sight of a person producing that sound. There is also evidence of a developmental change as early as 12 months in the ability to discriminate speech sounds. This change reflects exposure to the infant's native language.

How speech or sign perception interacts with syntactic and semantic knowledge in language comprehension is not understood to date. Research on the subject is now beginning, largely through the efforts and cooperation of many different kinds of scientists contributing a multi- and interdisciplinary perspective to this complex question.

Major Scientific Research Opportunities: Children

Normal Processes and Mechanisms

- Determine how sensory information from several modalities is used by

normal children in developing symbolic representation in language and literacy.

- Determine to what extent the integration of speech perception and speech production (motor) skills and psychosocial skills is an essential precursor to language development in normal children.

Deaf and Language Impaired Children

- Compare perceptual and motor processing patterns of language acquisition in deaf children acquiring spoken language and the language of signs.
- Develop appropriate *normative* and *criterion-referenced* psychosocial, intellectual, academic, and linguistic development appropriate for use with deaf and language impaired children.

Brain-Behavior Relationships

- Use imaging techniques and genetic studies to elucidate relationships between areas of brain functioning and language-related processes and mechanisms.
- Study the effects of gonadal hormones and the immune system on brain development in relation to language-related processes and mechanisms.

Major Scientific Research Opportunities: Adults

Brain-Language Relations

- Use refined neuroimaging techniques (both anatomical and metabolic) and electrophysiological techniques combined with analysis of functional impairment to determine how language is mediated by anatomical structures of the brain.
- Explore the role of various neurotransmitter systems in the mediation of particular language operations.

Behavioral Investigations of Language

- Determine the mechanisms involved in normal language processing in young adults and in the aging process, using theories and methods of cognitive science.
- Identify the processes underlying language disorders in patients with aphasia or dementia, including perceptual, motor, cognitive, and specifically linguistic mechanisms.
- Conduct comparative studies of different clinical populations to reveal common mechanisms and differences,

such as language disorders in hearing and deaf subjects, right- and left-brain-damaged subjects, language disorders subsequent to specific versus generalized brain damage, and language impairments in speakers of different languages.

Major Clinical Research Opportunities: Children

Understanding the Nature of Disorders and Atypical Language

- Identify and describe patterns of spoken language acquisition and acquisition of American Sign Language (ASL) in deaf children in relation to cognitive and psychosocial development, to language models provided (artificial or natural), and to perceptual and motor processes, and also when access to a first natural language is delayed.
- Develop the means for identifying language disorders in deaf children and normal hearing children from different cultures, and develop a rational typology for language impairments in affected children.
- Describe the perceptual/motor characteristics associated with different types of language impairment.
- Disambiguate cause and effect in children with both psychosocial disorders and language or language-based learning disorders.
- Look for common factors as revealed by linguistic profiles of language impaired children from different language-speaking communities.

Intervention Strategies

- Design and evaluate intervention strategies that:
  - are theoretically motivated;
  - are tailored to different subgroups of language or hearing impairment; and
  - include the use of assistive devices and of alternative (intact) modality sensory processing.
- Assess how best to involve parents in intervention with deaf and language impaired children.

Major Clinical Research Opportunities: Adults

Classification

- Improve the typology of language disorders through investigations of the linguistic, perceptual, motor, and cognitive mechanisms that are affected by brain damage.

Intervention Strategies and Recovery

- Develop intervention strategies for working with aphasic and brain-injured individuals, particularly strategies that capitalize on knowledge about normal language processing or that provide language impaired individuals with compensatory mechanisms, such as augmentative communication systems.
- Investigate effects of pharmacological agents upon the recovery process and on recovery augmented by clinical intervention in aphasia.

Major Scientific Research Opportunities: Speech and Sign Perception

Speech and Sign Perception in Infants and Children

- Investigate the ontogeny of speech perception, focusing on categorization, lipreading, and imitation in relation to critical periods and exposure to language.
- Study the relation of sign perception to speech perception with a focus on understanding whether language processing is similar regardless of the modality through which it is acquired.
- Relate early acquisition of phonology, assessed through tests of speech/sign perception, to the acquisition of higher levels of language such as acquisition of words (lexicon), word meaning (semantics), and grammar (syntax).

Speech and Sign Perception in Adults

- Characterize the basic properties of speech and sign—how speech and sign are generated, how they are perceived, how they vary cross-linguistically, and how the processes of speech and sign perception relate to those of speech and sign production respectively.
- Investigate basic and higher level processes underlying audition and vision, to aid in understanding the perception of speech and the perception of sign.
- Define how speech and sign perception relate to higher levels of language, including words (lexicon), word meaning (semantics), grammar (syntax), and discourse.
- Investigate how the perception of speech is affected by aging, and following hearing loss or brain damage.

Major Clinical Research Opportunities: Speech and Sign Perception

Assessment

- Standardize tests of speech perception in normal infants and young children to

assess the degree to which impairments of speech perception are predictive of language disorders.

- Develop methodologies for exploring speech perception and sign perception in adult patients with language disorders.

SMELL, TASTE, AND TOUCH

The chemical senses (olfaction and taste) and the tactile senses are the subject of chapter 6. The chemical senses play crucial roles in the detection of chemical stimuli in the environment, including foods, environmental chemicals and pollutants, hazardous vapors and gases, and pleasurable scents and fragrances. Deficits in these senses can eliminate the motivation to consume food and eradicate the ability to detect toxins in the home and workplace. Information about the prevalence and incidence of chemosensory disorders is difficult to assemble. In the late 1970s, the opinion of many experts was that more than two million adults in the United States had chemosensory problems, and patients were often given psychiatric referral or told that the problem would go away.

Even though extensive systematic data are not available, smell and taste disorders can affect diet, and marked changes in body weight occur in some individuals. Increases in intake of sweets, salt, and spices, to derive pleasure from eating, can exacerbate chronic health disorders such as obesity and hypertension, and losses in the capacity to identify certain odors can interfere with a person's ability to reject spoiled or contaminated foods or to identify "warning" odors such as mercaptan, which is added to natural gas. In addition, problems associated with a chemosensory disorder can be exacerbated when they are combined with a chronic illness such as cancer or renal or liver disease, contributing to anorexia and cachexia.

Research using modern neurobiological methods has led to the identification of odorant-binding proteins that apparently transport stimulus molecules to the olfactory receptor membrane. Although comparable proteins in saliva have yet to be described, scientists have found clues about the interaction between taste sensation and salivary secretion. Studies of central nervous system pathways have linked taste and smell with areas of the brain involved in drinking, feeding, swallowing, breathing, and sexual activity.

Studies in basic biology have led to the observation that there is early developmental plasticity in the chemosensory systems and that the opportunity for changing function and structure continues throughout life. In the gustatory system, cells that form the taste buds are replaced about every 10 days, and in some species, olfactory receptor neurons apparently undergo a cycle of turnover and replacement of about 30 days. These are the only known neurons with the capacity to replace themselves through life.

Improvements in diagnosis have taken different directions for taste and smell. In olfaction, a "scratch-and-sniff" booklet is

easy to use in a physician's office and can be distributed to large groups. Such identification tests are used to classify patients as anosmic (almost no ability to smell), hyposmic (reduced smell function), and normosmic (normal). In the area of taste, it is now known that about one-third of the population are "taste blind" to phenylthiocarbamide (PTC) and its chemical relative PROP. These "nontasters" are also insensitive to some other bitter and sweet compounds.

Perceptible olfactory losses appear to be more common than perceptible taste losses. Patients show a surprising inability to report localized taste loss. This appears to result from a compensatory mechanism. When one area of the tongue is damaged, other areas produce sensations that are more intense. This phenomenon maintains a relative constancy of taste experience. The primary etiologies associated with olfactory losses are head trauma, viral infection, and allergy.

There has been a growing appreciation of the devastating effects of chronic bad taste sensations (dysgeusia) and chronic bad smell sensations (parosmia). Improved diagnostic procedures have led to the conclusion that many cases are local in origin, such as postnasal drip. In addition, remarkable progress has also been made in the last decade in establishing the nature of chemosensory changes that occur over the life cycle. It is now known, for instance, that age takes a greater toll on olfaction than on taste.

Disorders of touch are often associated with direct trauma to the skin, such as frostbite, burn, penetrating wounds, and amputations, and with trauma to the central nervous system, such as stroke, concussion, congenital malformations, and perinatal anoxia. Estimates of the prevalence of disorders of touch range from 3 to 5 percent of the population. Studies during the past decade have resulted in the development of several tests of tactual sensitivity, which in combination with recently developed electrophysiological testing of single units of human peripheral nerves may prove to be an excellent tool in analyzing the extent of peripheral damage and course of recovery. For lesions of the central nervous system, tests of somatosensory function serve to delineate the area of damage as well as to track recovery.

Basic studies of tactile sensitivity have guided the development of aids for the disabled. Multichannel tactual aids, for instance, have shown considerable promise in improving lipreading performance. The use of these aids has led to information about some little-known sensory capabilities of the skin, and the aids are being used by scientists to answer basic questions about the sense of touch.

Major Scientific Research Opportunities: Smell and Taste

Plasticity, Including Development, Regeneration, and Aging

- Identify olfactory factors that control development of the brain.

- Use molecular probes as markers for specific taste bud cell types and developmental stages.
- Apply cell and tissue culture techniques to specify development of taste bud cells.
- Explore similarities and differences in development and maintenance of olfactory neurons, taste bud cells, and auditory hair cells.
- Explore the phenomenon of senescence in olfactory and taste systems to understand age-related chemosensory deficits.

Access of Infectious Agents and Toxins to the Brain Through the Olfactory Nerve

- Investigate the role of the olfactory nerve in the transport of toxins and pathogens to the brain.

Receptor Mechanisms and Signal Processing Throughout Central Systems

- Describe the access of odorant and tastant molecules to the receptor cells, including roles for mucus and saliva, and investigate the processes that could interfere with this access.
- Increase application of whole cell and channel recording techniques to augment the description of taste and olfactory transduction processes.
- Develop clonal preparations of taste and olfactory cell precursors to permit genetic and biochemical analysis of reception and synaptic transmission.
- Conduct studies to discern whether the central taste and olfactory systems are organized according to somatotopic and chemotopic principles.
- Characterize central neurotransmitters and neuromodulators with biochemical, pharmacological, and immunocytochemical techniques in olfaction and taste.
- Investigate changes in central taste and olfactory circuits associated with early development and aging.

Major Clinical Research Opportunities: Smell and Taste

Epidemiology of Smell and Taste Disorders

- Determine the incidence, prevalence, and risk factors of anosmia, hyposmia, parosmia, ageusia, hypogeusia, and dysgeusia, and observe the course of these disorders over time.
- Find the cause for disorders of chemosensory sensitivity in systemic diseases, and determine the prognosis for recovery.

Development of New Diagnostic Tests

- Develop additional diagnostic methods for smell and taste disorders as well as instruments for monitoring their course over time.
- Relate lingual and mucosal pathologies to smell and taste disorders.

Affective Dimensions of Taste and Smell

- Delineate the innate and acquired factors in chemosensory affect.
- Determine the role of odors in human social and sexual behavior and reproductive physiology.
- Investigate how smell and taste, in normal and deficient states, impact on food choice and utilization.

Associations Between Chemosensory Disorders and Altered Food Intake in Aging and in Various Chronic Diseases

- Assess the effect of the health status of the elderly on their chemosensory function.
- Investigate how chemosensory disorders impact on food choice in chronic diseases.

Effects of Environmental Agents on Chemosensory Function

- Assess the role of pollution and occupational exposure on chemosensory function.

Clinical Trials to Evaluate Potential Treatment Protocols and Rehabilitative Strategies

- Design and implement clinical trials of therapeutic strategies for sensorineural disorders of smell and taste.

Major Scientific Research Opportunities: Touch

Tactile Sensory Coding and Central Representation

- Investigate psychophysical and neurophysical processing of spatial/temporal information in the somatosensory system.

Touch and Its Interaction With Other Senses

- Study interaction among the cutaneous senses, including touch, temperature, the chemical senses, pain, and itch, at both psychophysical and neurophysiological levels.

- Study interaction between proprioceptive and cutaneous senses as well as between vision, hearing, and the cutaneous senses.

Major Clinical Research Opportunities: Touch

Development of Tactile Stimulators

- Develop more efficient and compact devices to present controlled, complex spatiotemporal patterns to the skin.

The Haptic Exploration of Objects and Surfaces

- Investigate individual differences in abilities for object identification by tactile exploration in normal and patient populations.

Tactile Sensory Aids

- Encourage research in tactile sensory aids, including speech-analyzing, reading, and mobility aids for deaf and deaf-blind persons.

Testing of Tactile Sensitivity in the Mouth

- Develop devices and techniques for testing tactile sensitivity in the oral-nasal cavity, and apply these to the population of patients with taste and olfactory complaints.

CROSS-CUTTING ISSUES

Chapter 7 is a discussion of trans-NIDCD concerns and program mechanisms. The chapter has six divisions: prevention and rehabilitation, specialized centers of research, data systems and population-based studies, research training and research personnel resources, the NIDCD national information clearinghouse, and multicultural research needs.

The division of prevention and rehabilitation discusses three types of prevention, along with approaches to intervention for rehabilitation: prosthetic, preventive, remedial, medical, and surgical.

The discussion of national research and training centers is in response to the mandate of P.L. 100-553 to develop, modernize, and operate research centers for studies of deafness and other communication disorders. The Task Force recommends that the NIDCD establish multidisciplinary National Research and Training Centers. Each center should have a central, unifying theme, combining basic and clinical research, and should strongly promote interactions among scientists across the research spectrum.

The discussion of data systems and population-based studies states that there

are no data on communication disorders at a national level, and highlights the need for expanded data systems. Better data are needed to study the incidence, prevalence, and risk factors for deafness and other communication disorders. Such studies should span the entire life cycle and include population samples reflective of the various ethnic and cultural backgrounds of the U.S. population. Recommendations in this discussion are also in response to a mandate of P.L. 100-553. The Task Force recommends that the NIDCD establish a Division of Epidemiology and form an advisory committee to it.

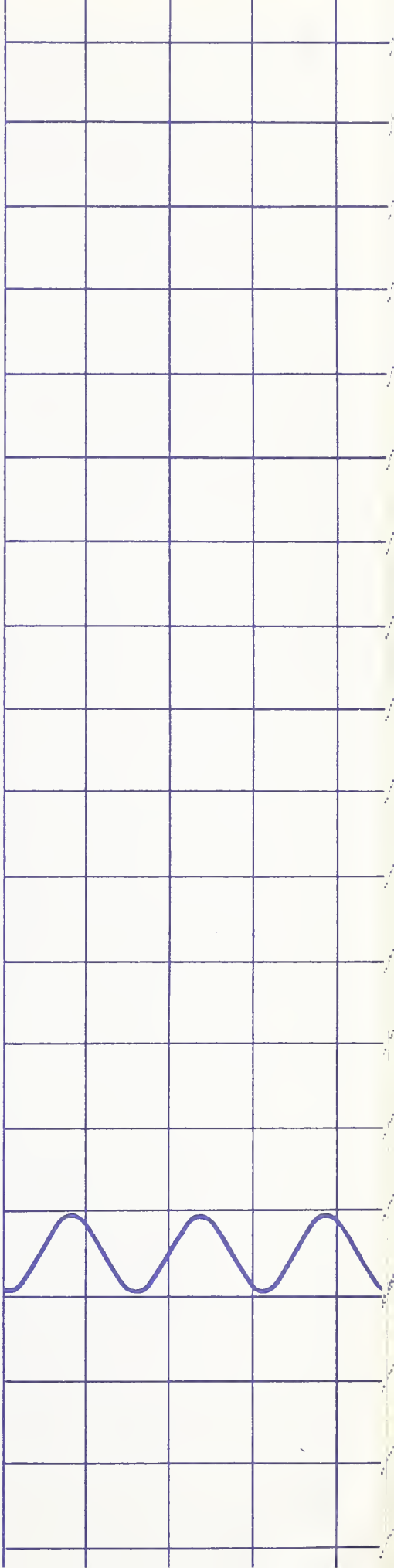
In the discussion of research training and research personnel resources, the Task Force recommends that a first priority of the NIDCD be to address a critical shortage of research personnel and training support. The dimensions of the problem should be carefully addressed, both quantitatively and qualitatively. When such information is available, the NIDCD should respond specifically and quickly to pressing needs in a cost-effective manner.

In the sixth division of chapter 7, the Task Force recommends that the NIDCD establish an advisory panel to plan an information clearinghouse, which is required by P.L. 100-553. The panel should define the scope of activity and develop guidelines for the NIDCD clearinghouse office.

The final division of this chapter draws into a single focus a concern reflected across scientific discussions in chapters 1 through 6. Although it is projected that one-third of the nation will be comprised of racial and ethnic minority groups by the turn of the century, only scant reference has been made to these groups in the literature on communication disorders. This discussion outlines needed approaches to the acquisition of epidemiologic data that are generalizable to specific populations.

1.

Deafness and Hearing Disorders



CONTENTS

OVERVIEW 39

Prevalence, Incidence, and Costs of Hearing Impairment 39
Historical Background 39
The Dynamic Cochlea 40
Relations to the Brain 41
Mechanisms of Complex Sound Perception 41
Localization of Sounds 42
Regeneration of Sensory Cells 42
Microbiology, Immunology, and Biochemistry of Otitis Media 42
Diagnosis of Hearing Disorders 43
Treatment and Rehabilitation of Hearing Loss 44
Environmentally Induced Hearing Loss 44

RECENT ACCOMPLISHMENTS 44

Normal Hearing 45
Hearing Disorders 46

PROGRAM GOALS 47

RESEARCH OPPORTUNITIES 49

Basic Hearing Sciences 49

Transduction Mechanisms in the Inner Ear 49
Functional Connections and Synaptic Mechanisms in the Auditory Centers of the Central Nervous System 50
Processing of Complex Sounds in the Central Nervous System 50
Perception of Auditory Space 51
Quantitative Systems Modeling of Auditory Function 51
Regeneration of Neurosensory Mechanisms 51
Development and Maturation of the Auditory System 52

Clinical Hearing Sciences 52

Role of the Immune System in Hearing Disorders 53
Molecular Genetics of Hearing Disorders 53
Otopathology 54
Auditory Consequences of Acquired Immune Deficiency Syndrome 54
Pathogenesis and Treatment of Tinnitus 55
Vaccine Development 55
Otitis Media 56
Clinical Trials in Sensorineural Hearing Loss 56
Clinical Trials of Cochlear Implants in Children 57
Epidemiology of Deafness 57
Identification of Congenital Hearing Impairments by Age of 9 Months 58
Loss of Hearing During Aging 58
Susceptibility to Environmentally Caused Deafness 58

Rehabilitation Devices and Behavioral Management of Hearing Loss 59

Comprehensive Understanding 59
Development of Neural Prostheses 60
Development of an Implantable Hearing Aid for the Middle Ear 60
Development of a Noise-Canceling Hearing Aid 61
Development of Tactile Aids 61
Research on Education for Hearing Impaired Children and Youth 61

Research on Prevention, Detection, and Diagnosis of Hearing Impairment 62

Children 62
The Elderly 63
Environmental Effects 64

SUMMARY OF RESEARCH RECOMMENDATIONS 66

Major Scientific Research Opportunities 66

Encoding of Sound by the Inner Ear 66
The Central Nervous System 67

Major Clinical Research Opportunities 67

Genetic Hearing Loss 67
The Bases and Rehabilitation of Severe and Profound Hearing Loss 67
Otology and Rehabilitation 68

1. Deafness and Hearing Disorders

OVERVIEW

Prevalence, Incidence, and Costs of Hearing Impairment

More than 28 million Americans are believed to have some degree of hearing impairment. This estimate includes approximately 2 million persons who are profoundly deaf. These numbers are expected to increase substantially in the next few decades due to increasing longevity and the consequent overall aging of the population.

One of every 1,000 infants is born totally deaf, and several times this number have a substantial impairment by the age of 3. More than one-third of the population has a significant hearing impairment by age 65, and the prevalence increases rapidly with increasing age. At least 15 percent of the population are affected by tinnitus, many so severely that it disrupts their lives. The socioeconomic impact of this epidemic of hearing impairment has never been rigorously estimated, but it is clearly vast.

The most common cause of temporary hearing loss is otitis media, which is predominantly a disease of infants and young children. Recent studies show that about 70 percent of all American children have had an episode of otitis media by the time they are 3 years old. This disease accounted for 10 million visits to doctors' offices in 1975 and an unknown number of prescriptions for antibiotics. The estimated health care costs for otitis media in the United States exceeds $1 billion per year.

Historical Background

Hearing impairment is invisible and nonfatal, two characteristics that attract little public attention. Research on hearing impairment has, therefore, lagged behind research on other less prevalent and less debilitating disorders. Also, a long history of societal discomfort and ignorance about deafness had to be overcome.

Historically, deafness has been linked both to mental illness and to mental retardation in the minds of an often unsympathetic, sometimes punitive public. In addition, well-meaning benefactors, teachers, and physicians of the deaf, in years now long past, resorted to crude and sometimes cruel interventions, alienating deaf people and politicizing options for their care. This legacy carries with it some unique "baggage" for contemporary deaf and hard-of-hearing people, the deafness research com-

munity, and the public, who now seek to expand research on hearing disorders.

Even now, there is insufficient public appreciation of the vital importance of good hearing to overall health and employment opportunities. All societies depend fundamentally upon the mutual abilities of their citizens to produce and understand speech and language. Hearing is integral to normal everyday human communication, and yet it is taken for granted much more than the rest of health. It is also still thoughtlessly and widely abused.

The absence of hearing produces isolation and a monumental barrier between the impaired individual and the hearing world. One consequence has been the evolution of a distinctive deaf culture, which lives within, but in many ways apart from, the hearing world inhabited by the majority of Americans.

In fact, many deaf individuals believe that deafness is not a disorder but a *culturally defining condition*. This group consists primarily, though not exclusively, of individuals who were born deaf or who became deaf during early childhood. Their distinguishing feature is that they use a sign language as a first language, even though some may also be proficient in spoken language. For many years, the special characteristics of this group were either ignored or even devalued by health and education professionals and researchers, whose principal concern was with hearing loss as a disorder. The NIDCD recognizes these important distinctions within the population of deaf persons.

Research on the ear and the processes associated with hearing and its disorders was inhibited for many years by the fact that the tiny sensory organ for hearing, the cochlea, is encased by bone and, unlike the eye, inaccessible to direct observation and experimental manipulation. The cochlea was, in fact, a "black box," which could be investigated in the living state only by indirect methods. Pathological studies were also difficult, since the preservation and histological preparation of the delicate structures of the cochlea were also complicated by the presence of the surrounding bone.

Some of these problems were circumvented, and substantial progress in research on deafness and hearing impairment began, most of it occurring within the past decade. Recent research on deafness has also benefitted greatly from major advances in biomedical research as a whole.

Fortunately, there have been significant changes in societal attitudes toward deafness, marked by a growing respect for deaf culture and an increased awareness of the prevalence and impact of hearing impairment. These changes have culminated in the landmark creation of the NIDCD within the National Institutes of Health (NIH).

Although the focus of the NIDCD Task Force was to identify, understand, treat, and ultimately *prevent* deafness and other communication disorders, the Task Force and the NIDCD respect the cultural integrity of deaf society and are committed to assist in breaking down barriers between the deaf and hearing worlds.

The Dynamic Cochlea

The inner ear not only receives but also *produces* acoustic energy signals, which are

called otacoustic emissions. This finding, about 10 years ago, opened a new era of investigation. Less than five years later, methods were found to isolate successfully and maintain outer sensory hair cells alive in vitro. This advance led to observations and investigations of hair cell movements in response to stimulation. Various experiments suggested that outer hair cell motility is influenced by the central nervous system (CNS).

These and related discoveries led to a revolution in the view of the cochlea. It is no longer conceived of as a strictly passive transducer of acoustic energy. Outer hair cells are the source of a mechanical feedback that greatly enhances the sensitivity and frequency selectivity of the inner ear. Moreover, disruption of this feedback leads to hearing loss, which is perceptually detected as an increased threshold for sound, a decreased range of sensitivity to sound, and a decreased frequency selectivity.

The origin of the unique fluid environment within the cochlea and details of intracochlear ion flow were established during the past decade, and the understanding of the biochemical mechanisms involved in transmembrane signalling is increasing. Intracellular recordings from inner and outer hair cells in vivo are now routine, and the recordings have revealed important functional differences between the two hair cell populations.

Relations to the Brain

Labeling techniques and electron microscopic examination have provided a precise anatomical description of the different afferent and efferent nerve fiber populations innervating the cochlea. For example, the efferent fibers innervating the inner hair cells receive input from a part of the brain that is different from the part that innervates the outer hair cells. Efferent nerve fiber activities appear to be important for the detection of signals in a noisy environment and may protect the ear from damage by loud sound.

Mechanisms of Complex Sound Perception

Complex sounds of everyday life consist of many spectral (frequency) and temporal components. It is known that the cochlea provides a specific nerve-impulse code for each of these acoustic components. Digital signal processing techniques implemented on small computers now allow scientists to generate, measure, and control extremely complex sounds, including speech. These techniques have led, in turn, to collaborative interdisciplinary studies of the representation of complex sounds across large populations of neurons and to investigations of the underlying neural mechanisms responsible for processing these representations.

Representation of a variety of speech sounds in the auditory nerve has been described in detail, and the representation of features of vowels in two cell populations in the cochlear nuclei in the lower brain stem has been elaborated. These findings have resulted from animal research. In the past three years, these observations have prompted the formulation of new models and mechanisms based on physiological and

psychoacoustic data to account for the perception of complex sounds, such as speech.

Localization of Sounds

Among the most exciting of recent advances has been the elucidation of neural mechanisms responsible for specific auditory behaviors in animals. For example, the representation and processing of target-identifying features of bat sonar signals in the auditory part of the brain's cortex have been elegantly described, and a neural map of auditory space has been constructed for the barn owl. This work is adding significant new knowledge to the understanding of human sound localization.

Regeneration of Sensory Cells

Decades ago, it was shown that production of sensory cells in the ears of mammals ceases before birth. This finding confirmed that sensory cells are produced only during embryonic development (only an assumption previously), and meant that damage to sensory cells later in life was irreparable. This fact is the basis for considering "nerve" deafness a permanent and irreversible condition.

It is now known that sensory cells are continually being added to the functional populations of cells in the ears of cold-blooded animals. Even more recently, it was shown that such sensory cells can be regenerated on demand in damaged cochlear epithelia, even in species where production of these cells normally ceases early in embryonic development. This regenerative potential has been confirmed in several species and is not limited to immature animals.

These completely unpredicted results from basic investigations of ear development in cold-blooded animals have provided an opening to understanding and perhaps manipulating the growth of human sensory cells.

Microbiology, Immunology, and Biochemistry of Otitis Media

Recent advances in microbiology include a better understanding of how respiratory viral infections contribute to the pathogenesis of otitis media. In addition, more is now known about the role of certain bacterial cell surface components, particularly endotoxin, which remain in the middle ear long after bacteria are killed and cause a sustained inflammatory response leading to persistent fluid in the middle ear. This is an important mechanism responsible for the development of chronic otitis media with effusion.

The role of the immune system in susceptibility to and recovery from otitis media has been extensively investigated, revealing that an IgG_2 subclass deficiency is an important susceptibility factor for otitis media. This finding has led to a better understanding of middle ear immune mechanisms and their genetic regulation.

Even more is known about the natural nonimmunological protective mechanisms in the middle ear, such as naturally occurring

lysozyme and lactoferrin. The protective role of the mucociliary system in the eustachian tube has been recognized, in addition to the tube's muscular function in protection. Nearly all individuals with impaired tubal mucociliary systems suffer from persistent or recurrent otitis media and sinusitis.

Diagnosis of Hearing Disorders

Quantum leaps have been made in the development and use of noninvasive diagnostic procedures. Three major advances in diagnostic capabilities are based on immittance audiometry, auditory brain stem response (ABR), and insertion gain measurements.

Immittance audiometry has permitted more exact diagnoses of middle ear disease. Measurement of the middle ear muscle reflexes has increased the likelihood of accurately identifying the site of a lesion and provided insight into how the ear is able to process speech at high intensity levels.

The widespread use of ABR tests has led to major advances in assessing the integrity of the auditory pathway in the brain. This noninvasive procedure provides valuable information about how well certain portions of the auditory system in the brain respond to a presented stimulus. The technique has proven most useful in the early detection of hearing loss in newborns and infants, for confirming nonorganic hearing loss, and for detecting tumors or other diseases affecting auditory function in the brain.

Development of new stimulus delivery systems and recording electrodes now permit use of ABR for intraoperative monitoring and monitoring of patients with head injuries. Electrodes can now be comfortably positioned on or near the ear drum for routine use in electrocochleography (ECoG), which may be helpful in diagnosing Meniere's disease. Another technique is the measurement of otacoustic emissions, which are sound signals produced by the inner ear, sometimes spontaneously, and sometimes triggered by an incoming sound. This technique shows great promise as a quick and reliable screening device. It may be particularly useful in the early identification of hearing loss in infants.

Hearing loss in many elderly patients is progressive, starting first at the higher frequencies and gradually progressing to lower and lower frequencies. The development of devices for testing perception of high-frequency (high-pitched) sound is another active area of research.

Identifying auditory dysfunctions within the central nervous system remains an imperfect art, particularly when unrelated inner ear dysfunction is also present. A number of new diagnostic techniques under investigation may prove useful in this area. Chief among them are magnetic resonance imaging (MRI) and positron emission tomography (PET). Current research is focused on determining whether these techniques are precise and selective enough to detect at least gross abnormalities in the function of various auditory centers within the CNS, even when structures appear

normal. ABR combined with modern computer signal processing techniques may become even more useful in this regard.

Bilateral conductive hearing loss has historically been difficult to diagnose because of the masking dilemma it creates. This problem was alleviated by the advent of insert ear phones.

Treatment and Rehabilitation of Hearing Loss

The simultaneous expansion of knowledge in such areas as cochlear physiology, computer technology, and miniaturization of electronic circuits has led to the development of greatly improved assistive devices. The most dramatic of these devices is the cochlear implant, several types of which are currently in clinical use. While much remains to be learned about their use and long-term effects, most implant recipients now perceive some sounds and, in some instances, recognize speech without the aid of speechreading.

Hearing aid evaluations have also become more objective with the development of insertion gain measurement equipment. This procedure provides the audiologist with an objective measure of the amount of amplification of the hearing aid for each frequency. The result is a more appropriate match to the specific hearing loss of a patient, without introducing overamplification.

Environmentally Induced Hearing Loss

During the last decade significant progress has been made in understanding how hearing loss may be caused by environmental factors such as noise, drugs, and toxins. For the first time, rational hypotheses have been advanced for the mechanisms underlying hearing loss induced by aminoglycoside antibiotics, leading the way to the design of protective measures and drug modification. The understanding of inner ear damage by diuretics has reached the point that preventive pharmacological treatment has been successful in animal models.

The way noise affects the inner ear is better understood. Advances include novel and important information about the effects of noise on cochlear microcirculation, fluid homeostasis, and mechanical properties of the sensory cells. These advances provide an excellent basis for future research to elucidate the molecular mechanisms underlying environmentally induced hearing deficits, and they strengthen the hope of finding means to prevent or ameliorate them.

RECENT ACCOMPLISHMENTS

Both basic and clinical investigators in the deafness research community are actively undertaking research in many new directions. They are exploiting past accomplishments and taking advantage of the tools

and insights offered by recent major progress in neuroscience, cell biology, immunology, molecular biology, genetics, and computer technology. Progress in understanding the mechanisms of normal hearing is accelerating. Comparable progress in transferring new knowledge into clinical applications, however, is not occurring.

The relatively slow rate of applying new knowledge to the problems of patients is due, in part, to normal delays. Part of the delay in clinical application, however, must also be attributed to a severe shortage of clinical investigators who are conversant with the state of basic hearing research and with the many emerging opportunities for new and improved clinical procedures. The gap between bench research on deafness and clinical care has widened to an alarming degree, as discussed in chapter 7, "Research Training and Research Personnel Resources."

Normal Hearing

Within the past year, many insights into the function of the cochlea have been derived from in vivo and in vitro studies. Considerable progress is being reported in measuring cochlear blood flow and studying its regulation. Studies of cochlear endolymph are providing insights into the regulation of the properties of this unique fluid as well as the effects of these properties on transduction. The immunobiology of the inner ear has been extensively characterized. It has been possible to show that changes in hearing are associated with the loss of outer hair cells. More has been learned about stereocilia; they are physically linked at their tips, and they participate in frequency transduction, or "tuning." It is also now clear that the frequency map of the cochlea is not a constant. It can be reprogrammed.

In vitro studies of isolated hair cells have led to a greater understanding of the biophysics of mechanoelectrical transduction and of ion channels in hair cell membranes. Progress has been made in the molecular and functional characterization of the structural and contractile proteins of stereocilia, hair cells, and the cuticular plate. This knowledge contributes to a better understanding of how stereociliary stiffness is modulated. Biochemical studies have revealed the presence of second messenger molecules in outer hair cells, which may play a role in their slow motile mechanisms. Modern immunohistochemical techniques have shown a co-localization of neurotransmitters at hair cell synapses and are being used to identify the proteins associated with ion channels. Species differences in hair cell membrane resonances have been found. Ultrastructural studies have revealed details about the contact zones between outer hair cells and efferent nerve fibers. In addition, cells and tissues from the inner ear can be grown in tissue culture, and hence are available for in vitro studies.

Auditory nerve fibers connecting hair cells to cochlear nucleus cells in the brain stem have been characterized with new intracellular marking techniques. The membrane properties of these cells have been analyzed with in vitro techniques that allow manipulation of the electrical and chemical environments of the cells. Connections between certain cell types in one brain

auditory structure and another have been established through tracing with markers that are specifically indicative of different types of neurotransmitters. An emerging technique of vast importance in neuroscience is multiple electrode recording from a number of brain cells simultaneously. A multi-electrode array has been developed for recording from cells within the parts of the brain associated with the auditory system. Medial and lateral olivo-cochlear bodies and their transmitters were shown to have multiple origins. Changes in the central nervous system in response to cochlear destruction and/or deprivation of auditory nerve signals have been described. There has been substantial additional progress in understanding the encoding of complex signals transmitted from the auditory nerve to the cochlear nuclei in the brain stem. The relationship between the neural code for sound intensity frequency and temporal structure, and the perception of these stimulus variables have been further clarified.

Major new approaches have emerged for studying auditory localization. By measuring the transfer of acoustic information between a sound source and the sound arriving at the ear drum, sounds can be reprocessed backward and presented over head phones in such a way that the sounds are perceived as if they had occurred in real auditory space. These techniques are providing an economical and efficient means for studying localization. They are demonstrating the relative importance of head and torso position for localization, and are providing new possibilities for presenting sounds in simulated situations, such as over hearing aids.

The progenitors of regenerated sensory cells may be the key to the self-healing of the ear's sensory epithelium, just as the cells that produce new skin are the key to wound healing. The progenitor cells of a cold-blooded animal's sensory cells have been identified and are the focus of intense investigations evaluating the molecular mechanisms involved, as well as of treatments that may be used to trigger and to control this healing process.

Hearing Disorders

Research on the molecular genetics of hearing disorders in mutant mice has shown positive correlations between changes in structure and function of the auditory system. Genetic analysis of selected deaf families is under way. Immunological markers have been identified that are predictive of genetic risk of developing chronic otitis media. Recent epidemiological studies have indicated a high prevalence of otitis media and shown that a large number of the persons affected had no clinical symptoms and normally would have gone undetected. Investigators are beginning to characterize cell membrane receptors for major pathogenic organisms causing otitis media. New evidence suggests that the middle ear immune response can be manipulated by donor T cells from animals presensitized by oral antigen. Studies have shown that bacterial toxin present in the middle ear cavity can penetrate the round window membrane and enter the inner ear, where it causes endolymphatic hydrops and

sensory damage. The inflammatory mediators involved in various types of otitis media have been characterized biochemically.

On the horizon are exciting new applications of monoclonal antibody techniques for research on hearing disorders. These markers could eventually assist in the diagnosis of hearing disorders by discriminating between normal and abnormal cell populations within the auditory system. Moreover, toxins can be joined to such antibodies, possibly providing a nonsurgical means for eliminating abnormal cell populations, such as acoustic tumors. This technique is being developed as a cancer treatment in other organ systems.

Intensive research has continued on cochlear prostheses, and additional progress has been made in new speech coding strategies. Ongoing evaluations indicate that different types of electrical stimulation of a given device can produce very different degrees of success in perceiving speech.

Cochlear implants may provide little benefit for subjects who have very few surviving cochlear nerve fibers. Methods for assessing the number of nerve fibers remaining in an individual candidate for cochlear implantation are greatly needed. This is an area of intense ongoing investigation. For patients who have extensive nerve damage and are not candidates for cochlear implants, neural prostheses implanted in various auditory centers of the CNS may be a solution. Research directed toward developing prostheses for use in the CNS is under way.

A real threshold in the design of hearing aids was crossed during the past year with the commercial introduction of a completely digital (computerized) hearing aid.

Sensorineural hearing loss is currently treated with behind-the-ear, canal-type, and in-the-ear (concha) hearing aids. All of these aids have inherent problems, some of which should be alleviated by implantable hearing aids that are capable of directly driving the middle ear bones (ossicles).

PROGRAM GOALS

Research to alleviate deafness has made measurable strides in the past several years and, under the auspices of the NIDCD, promises to provide even more insight into hearing and, more importantly, into how one may better treat hearing disorders and deafness.

The prevention, diagnosis, treatment, and rehabilitation of hearing disorders depends upon the identification of the disease process, an understanding of its underlying basic mechanism, and the development of effective strategies of intervention and rehabilitation. Success in these endeavors requires a close integration of clinical and basic research.

An increasing number of new evolving technologies will be applied to the elucidation of normal hearing processes, particularly those concerned with how speech and other biologically meaningful sounds are understood. Major progress is expected in learning how disruption of these normal processes may result in the numerous types

of known hearing disorders. This will allow detection of new classes of disorders as well as earlier detection.

Further understanding of the mechanisms of normal hearing and hearing disorders rests upon the synthesis of numerous factors: from understanding the physics of sound and the ear through identifying molecular events at the cellular and subcellular level, to analysis of the neurological processing of sound information in the nervous system, to learning how these activities and interactions result in the perception of sound, and to understanding auditory perception.

A multidisciplinary approach to the study of hearing is required to interpret these complex events. In fact, recent rapid increases in the knowledge of the hearing process have already resulted from the integration of strategies and technologies of a wide variety of fields. Unfortunately, the clinical application of this new knowledge has been slow.

Progress in key areas offers special opportunities to enhance diagnosis, treatment, and prevention of many of the disorders that cause hearing loss and deafness. Pursuit of these opportunities will significantly reduce the incidence and prevalence of hearing loss by the end of the next decade.

Consideration of these factors leads the Task Force to recommend that the NIDCD plan a comprehensive program of research with the following special characteristics: explicit acknowledgment of the multidisciplinary base of the field, accelerated integration of basic knowledge with clinical needs, and expeditious implementation of the mandate of Public Law 100-553. The broad objectives of the deafness and hearing disorders program include the study of normal mechanisms as well as the clinical disorders that disrupt the auditory function, and include the following:

- Goals of studies of normal hearing processes
  - Determine how the ear processes and encodes sound.
  - Determine how the CNS processes the output from the ear.
  - Determine how CNS processing leads to perception and behavior.
  - Characterize the life cycle (development, maturation, and aging) of the normal auditory system.
- Goals of studies pertaining to clinical needs
  - Develop population studies to determine and track the incidence, prevalence, and risk factors of hearing loss.
  - Initiate multidisciplinary studies to detect and determine the pathogenesis of specific hearing disorders.
  - Conduct clinical trials to establish the efficacy of existing and new treatments of disorders that cause hearing loss.
  - Develop and improve devices and rehabilitative strategies to assist hearing impaired people.
  - Develop scientifically sound medical and public educational strategies to prevent hearing loss.

RESEARCH OPPORTUNITIES

Details of a comprehensive plan for intramural and extramural research are found in appendix E. Specific strategies that should receive priority in both basic and clinical hearing sciences are outlined below.

Basic Hearing Sciences

The understanding of any process of disease or impairment requires basic knowledge about how the affected system functions in its normal state. In the search for this knowledge, auditory scientists have recently made some exciting fundamental discoveries. Exploitation of these new advances during the next few years promises to provide essential new information about how the auditory systems of humans and animals are organized and how they function. Areas that offer particularly strong opportunities for progress are discussed below.

Transduction Mechanisms in the Inner Ear

Over the past decade, progress has been breathtaking in understanding the transduction of sound into a nerve-impulse code in the inner ear. The discovery of acoustic energy originating in the inner ear (otacoustic emissions) and of stimulus-evoked motility of outer hair cells has led to a revolution in views of how the inner ear functions and also to novel experimental approaches. The results promise more progress in the next few years in basic understanding and clinical applicability.

This characterization of inner ear mechanisms will be greatly facilitated by the continued application of new methodological breakthroughs such as modern optical imaging and measuring techniques and the use of hair cells in culture. The research momentum now existing must be maintained to achieve significant results. Research in the following areas appears especially promising:

- The control of mechanical vibrations in the cochlea.
- The mechanisms of the transduction of mechanical vibrations into nerve impulses.
- The processes that maintain a constant environment in the inner ear (homeostatic mechanisms).

Studies in these areas are fundamental to understanding auditory system function and to developing improved rehabilitation techniques, such as better hearing aids and speech processors for cochlear implant devices. An expansion of basic knowledge about homeostatic mechanisms, which concerns blood flow and other metabolic features of the inner ear, is essential for understanding the pathogenesis and potential treatments of many forms of sensorineural hearing loss, including presbyacusis.

Functional Connections and Synaptic Mechanisms in the Auditory Centers of the Central Nervous System

The nerve-impulse code generated in the inner ear and transmitted along the auditory nerve is processed by brain networks of neurons, which are connected with synapses. A synapse can transmit either an excitatory or an inhibitory influence from one neuron to another. The responses of a neuron (and hence its signal processing functions) are determined by the pattern of synaptic inputs it receives.

In the past 10 years, an explosion has occurred in techniques for the study of signal processing in individual neurons at the level of their cell membranes and associated synapses, as well as techniques that have allowed the definition of specific circuits. Exciting progress is being made toward the goal of detailed descriptions of the mechanisms of signal processing in several identified types of neurons in the CNS auditory centers. For the first time, investigators are on the threshold of understanding the components of the complex neural networks of the auditory system.

High priority should be given to studies of synaptic mechanisms and functional connections of neurons in the auditory system. These studies should employ the wide range of existing and emerging techniques of modern neuroscience, including tissue slice and isolated cell preparations, tracing techniques, intracellular staining to identify precisely the neurons under investigation, and the use of multi-electrode recording and physiologically detailed computer models of neural organization.

Processing of Complex Sounds in the Central Nervous System

Most sounds of biologic importance consist of many temporal and spectral (frequency) components. Much is known about how the nerve-impulse codes for these components are generated in the inner ear. Relatively little is known about how these codes are processed by the auditory centers in the CNS to form the perceptions associated with the sounds that are heard. Recent physiological and perceptual research has revealed important information about interactions among the components of complex sounds. New studies are needed to develop a unified understanding of how the interactions are treated by the nervous system. The studies can use the technologies that relate physiological findings to new information about CNS connectors and to a new psychophysical understanding of the interactions inherent in complex sounds. New technologies permit the simultaneous study of nearby single cells of the central nervous system and also permit simultaneous study of neurophysiological activity of single cells in animals that are awake, in which perceptions can be measured. Findings from such studies are leading to entirely new ways of thinking about auditory perception and paving the way for fitting individual neurons and groups of neurons into the CNS processing scheme for complex sounds.

Perception of Auditory Space

Basic to the survival and evolutionary success of a species is its perception of auditory space and its abilities to find mates and avoid prey. Recent advances in understanding the acoustic transformations of sound provided by the head and external ear and their consequences for psychophysical measures of localization, and binaural (both ear) processing of sounds in auditory centers within the central nervous system, together with progress in analyzing the specialized systems of animals such as owls and bats, should lead to greatly improved knowledge of how humans determine the location in space of the sources of sound. This ability not only allows humans to identify potential dangers; it also relates to their ability to hear and converse in noisy environments, such as parties or sports events. This ability is first compromised with many forms of hearing loss. As knowledge of these phenomena increases, it should be possible to improve the ability of hearing impaired people to hear in noisy environments, to improve the naturalness of sounds reproduced by ear phones and hearing aids, and to create better interfaces between humans and computers in robotics and other sensing applications.

Quantitative Systems Modeling of Auditory Function

Rapid advances in understanding the auditory mechanisms in the inner ear and CNS permit the generation of quantitative models for auditory function that are based on physiological and psychophysical data. Many of these models have become feasible only with the advent of large-scale computer facilities. Specific examples of systems to be modeled include macro- and micromechanics of the inner ear; mechanical, electrical, and electromechanical transduction by hair cells; active processes of neural control of transduction events; representation of speechlike stimuli in trains of auditory nerve-impulse code; neural processing of auditory nerve signals by cell groups in the CNS; sound localization; and relationships between auditory nerve signals and human auditory perception. These models are essential; they allow for making predictions and forming hypotheses that can be tested experimentally. Such models are also applicable to the design of coding strategies for prosthetic devices and could contribute to the development of computer "neural networks" simulating and perhaps augmenting brain function.

Regeneration of Neurosensory Mechanisms

Sensorineural or "nerve" deafness has been considered irreversible because the production of the permanent sensory and nerve cells of the ear normally ceases before birth (in the fetus). Recent basic research with animals, however, has shown that under certain conditions sensory cell production can be reactivated in mature damaged ears. It is also known that these regenerated cells contribute to a recovery of hearing. Study of the cellular mechanisms and the molecular control of this self-repair process is readily amenable to methods of

modern biotechnology. Basic elucidation of the process should lead to therapeutic advances.

It is reasonable to expect that within the foreseeable future the progenitors of regenerated sensory cells will be cloned from animals and that the product will become available for the development of pharmacological agents for controlling regenerative cell replacement and auditory recovery. In addition, the natural molecular triggers for sensory cell growth and inhibition and the potential occurrence of spontaneous sensory cell regeneration in species more closely related to humans should be extensively evaluated.

The realization of clinical gains from this research is likely to require 10 to 20 years of sustained investigation, but the potential benefits are great. For 80 percent of the 28 million Americans affected by hearing impairment, their loss is currently irreversible, due to inner ear deafness. The studies proposed here suggest that this type of deafness might not have to be considered permanent.

Development and Maturation of the Auditory System

The development and maturation of normal hearing and the auditory problems of children are now appropriate subjects for systematic and comprehensive investigation. Investigators should continue to gain insights into the auditory abilities of infants and young children. It is also necessary to determine, with animal models, the molecular genetic basis of normal and abnormal auditory structure and function. Research on embryonic mechanisms in the formation of the normal ear and also of normal, functioning auditory pathways in the brain will lead to an understanding of mechanisms that produce congenital defects and an ability to correct them. In addition, understanding the normal developmental changes in plasticity within the auditory system will have importance for both the periphery and the brain. The importance of critical periods for functional development as well as changes in the strength of interactions and interdependencies in the auditory system are likely to be related to changes in susceptibility to a compromise of hearing function. In addition, variable influences of environmental factors through several stages of auditory development should be evaluated. Metabolic, biochemical, and immunological changes in the middle ear, the cochlea, and the brain over the life cycle of the normal auditory system should be investigated.

This research should be integrated with studies focused on characterizing the cellular basis and the mechanisms that regulate sensory cell regeneration and recovery of hearing in damaged auditory systems, both in animals and in tissues and cells maintained in culture.

Clinical Hearing Sciences

Recently, a rapid increase in understanding the basic mechanisms of normal hearing and some disease processes has occurred. The clinical application of this new knowledge, however, has been slow, in part because of the shortage of clinical

investigators. Application of basic research results in several key areas will enable a substantial reduction in hearing loss in the next decade. Areas that offer particularly strong opportunities for progress in clinical care are discussed below.

Role of the Immune System in Hearing Disorders

The immune system is crucial to the defense of both the middle and inner ear against infection. In recent years, the role of immunological mechanisms in many disorders of hearing has been increasingly recognized. Inflammation generated by the immune response undoubtedly contributes to tissue damage associated with infection. Evidence suggests that immune mechanisms also induce hearing loss and are responsible for the progression of chronic otitis media. Immune mechanisms may also contribute to the pathogenesis of other disorders for which no cause has been established, such as Meniere's disease.

Research is needed on normal immunological activities in the middle and inner ears as well as on the immunological response to infecting organisms. Animal models of autoimmune sensorineural hearing loss should be developed, if possible, for research on the pathogenesis of this phenomenon. Improvements in methods for diagnosing autoimmune hearing loss are also greatly needed. Application of research techniques of neuroimmunology to the auditory system should contribute to a rapid expansion of knowledge in this evolving field.

Molecular Genetics of Hearing Disorders

Genetic factors are known to cause over 50 percent of all cases of severe childhood deafness. The impact of heredity upon late onset hearing disorders is mostly unknown, but it is believed to be considerable. Despite the significant role of genetic factors in causing hearing impairment or deafness, little research has been conducted in this area. Significant progress has been made in mapping the genes of the few X-linked disorders involving hearing loss (Norries disease, perilymphatic gusher, and Alport syndrome). Recent advances in genetics and molecular biology have opened an opportunity for research that should lead to an understanding of how hearing impairment and deafness are transmitted from parent to offspring. Such studies should also do much to clarify the biology of hearing and the molecular nature of its disorders.

The mapping of genes on chromosomes that cause deafness or other types of hearing impairment and the characterization of such genes and their expressed products (enzymes and other proteins) should be given a high priority. This objective can be achieved through the study of large families with members with inherited deafness, by the construction of DNA libraries for auditory structures in the human inner ear and brain, and also through study of animal models of genetic deafness. These and other molecular genetic techniques will lead to the cloning and decoding of genes that cause deafness. Means of earlier diagnosis and more precise methods of genetic counseling and medical management can then be

developed. Ultimately, genetic engineering techniques may allow correction of hearing impairments.

The approach to the study of the genetics of hearing loss should be multifaceted. A variety of known disorders should be targeted for early study. This should include such disorders as Waardenburg syndrome, Usher syndrome, otosclerosis, adult onset hearing loss, presbyacusis, and otitis media.

Otopathology

With the rapid advancement of hearing test methodology and immunological and molecular biological techniques, opportunities now exist to develop a knowledge of ear pathologies at a more sophisticated level. Collection of human temporal bones, which was previously funded largely by the NIH, must be resumed. In the past, such collections provided valuable descriptive information about human ear pathologies. There is also a critical need to collect human tissues of the auditory system (both temporal bones *and* auditory portions of brains) from individuals with a variety of well-documented deafness-causing disorders and to correlate the pathology with advanced genetic, audiometric, neurophysiological, and psychoacoustic measurements. Blood/serum and skin should be collected from these same individuals for future use in immunological and genetic studies.

These efforts should be complemented by normative histological data on the auditory system in relation to age, race, and sex in order to provide a basis for assessing age-related morphologic changes and to interpret pathological effects on the central auditory system. Histopathological studies should be extended to include modern methods of tissue preparation, which allow comprehensive evaluation with immunohistochemical, molecular biological, and ultrastructural investigations.

Methods should be established for storing data in a data bank that can be shared by investigators and institutions.

Auditory Consequences of Acquired Immune Deficiency Syndrome

Infection with human immunodeficiency virus (HIV) can result in fully developed acquired immune deficiency syndrome (AIDS), characterized by immunosuppression, the presence of Kaposi's sarcoma, and a high level of opportunistic infections. HIV infection can also lead to AIDS-related complex (ARC), which is similar to AIDS but has a lower level of opportunistic infections. Both AIDS and ARC are associated with neurological complications that include auditory impairments. It is estimated that 75 percent of adult AIDS patients and 50 percent of ARC patients show clinical auditory system abnormalities.

The etiology of auditory system changes in HIV patients is unknown. Direct infection of the nervous system by HIV is well documented, and strains of the virus are known to selectively infect specific neural populations. Hearing disorders in AIDS patients could thus be caused directly by preferential HIV infection of the cochlea or the central auditory system. Many of the

complications of AIDS, however, are the result of opportunistic infections rather than of HIV itself. By far the most prevalent cofactor is cytomegalovirus (CMV), with more than 90 percent of AIDS patients showing active CMV infections. Since CMV is known to preferentially damage the auditory system in congenital infections, it seems likely that CMV is also involved in HIV-related auditory pathologies as well. Research to clarify the role of HIV and CMV infection in the auditory pathologies of AIDS patients should lead to improved strategies for treating the hearing deficits that are so common in this population.

Pathogenesis and Treatment of Tinnitus

Many persons with ear disorders hear noises (ringing, buzzing, or roaring) when no external acoustic stimulus is present. This disorder is called tinnitus. At least 15 percent of the American population have frequent or constant tinnitus, and 10 percent of these people have tinnitus that is severe enough to disrupt their lives. Persons over 50 are twice as likely to have tinnitus. Research on tinnitus is in its infancy and should be greatly expanded.

The natural history of tinnitus resulting from trauma or disease is unknown. While some data on the prevalence of tinnitus in adult populations are available, the types and degrees of disability caused by tinnitus need to be characterized and measured in population surveys.

Aspirin toxicity causes tinnitus, while simultaneously affecting inner ear hair cell motility in a way that can be measured noninvasively. A possible relationship between otacoustic emissions and some forms of tinnitus deserves additional study. Animal models can help elucidate mechanisms and sites of origin of tinnitus, and can also be used to test clinical treatments such as electrical suppression and acoustic masking. Such models promise new insights into both tinnitus and normal inner ear function.

Reliable and valid clinical measures of tinnitus sensation and disability must be developed for use as outcome measures in future studies of existing and new treatments.

Vaccine Development

A major priority for the NIDCD is to help develop vaccines that would significantly reduce the incidence of infections causing meningitis, which may lead to deafness, or causing otitis media, which may lead to temporary or fluctuating hearing loss.

Field trials have proven that multivalent pneumococcal capsular polysaccharide vaccine is largely ineffective for the very young, who are in the greatest need for protection because of the immaturity of their immune systems. With the advent of molecular biological techniques and a deeper understanding of protective immune mechanisms, it should be feasible to develop new types of vaccines, including conjugated vaccines against bacteria causing otitis media. These new vaccines would contain a carrier molecule attached to the bacterial component, which acts to enhance the immune response of very young children.

Conjugated vaccines against *Hemophilus influenza* type b, which is the major common pathogen causing meningitis, are in varying stages of development and require field tests and clinical trials. If proven effective, such vaccines would make a significant contribution to preventing deafness.

Otitis Media

Otitis media is an inflammation of the middle ear and is classified into a spectrum of disease types. Some children are prone to recurrent disease, and their risk factors are poorly understood. Surgical management consists of inserting small tubes through the ear drum to ventilate the ear. Medical management includes administration of antibiotics, antihistamines, decongestants, mucolytic agents, and anti-inflammatory agents. Definitive studies regarding the efficacy of these various treatments alone and in combination are lacking.

Strains of bacteria that commonly cause otitis media tend to acquire resistance to antibiotics, but most patients are treated empirically without any sampling and culturing of middle ear fluids for bacteria. There are no standardized data addressing the relationship of age, sex, race, anatomic, and environmental factors and the effect of antibiotic and other forms of medical and surgical treatment.

There is growing evidence suggesting that repeated episodes of otitis-media-induced hearing loss are associated with vocabulary deficits, language learning deficits, aberrant behavioral patterns, and reduced educational achievement. There is therefore a very serious need to expand research on the risk factors, diagnosis, treatment, and consequences of otitis media, including clinical trials of treatment modalities.

Genetic markers of the immune system should be investigated in an effort to identify high-risk populations, which could then be targeted for intensive preventive measures. There is evidence that the epidemiology of otitis media varies across racial groups. Such differences appear to be linked to variations in the anatomical and structural characteristics of the eustachian tube. Expansion of comparative group research in this area would provide further clues to the genetic etiology of otitis media.

The advisability of impedance screening for preschool and school-age children needs to be assessed. Longitudinal studies are needed to determine the relationship between otitis media and delays in child development.

Clinical Trials in Sensorineural Hearing Loss

For sensorineural hearing losses that are sudden, fluctuant, or rapidly progressive, a variety of empirical therapies are currently used. Often these therapies are based on assumptions about the pathogenesis of the hearing loss. Fluctuating losses, for example, are often believed to be due to endolymphatic hydrops and are treated with diuretics and salt restrictions, while losses believed to be due to inflammation or ischemia may be treated with steroids or vasodilating agents, respectively. There is some evidence supporting the therapeutic

efficacy of steroids in certain patients with sudden hearing losses; in general, however, medical therapy for sensorineural hearing loss has not been shown to be efficacious.

There is a pressing need for properly controlled therapeutic trials in these areas. The use of agents affecting cochlear blood flow, for example, needs to be studied both in sudden and in rapidly progressive sensorineural hearing loss as well as in fluctuant hearing loss. Studies comparing bed rest, carbogen breathing, steroids, and hemodilution also need to be conducted.

Some of the treatments now being used empirically may be effective only in certain subsets of patients. If there were a better way to identify such patients, treatment of them would be more rational, and effective treatments could be more readily identified. Further studies with animal models of sensorineural hearing loss are essential to provide insights into pathogenesis and to develop diagnostic tests that will permit clinicians to identify pathogenetic factors in their patients.

Clinical Trials of Cochlear Implants in Children

Cochlear implants have significantly improved the communication skills of postlingually deafened adult implantees. Recently this technology has been extended to pre- and postlingually deafened children. Initial results with a single-channel implant suggest that deaf children can achieve some awareness of sound with this device. In the past year, reports suggest that some congenitally deaf children have obtained some word understanding following implantation with a more advanced multichannel implant system.

Extensive clinical research is necessary to address the impact of this technology in children and to establish its efficacy. This research should assess the audiological and psychophysical performance, speech and language development, and educational progress of these children. In addition, studies should be initiated to determine the most appropriate method to integrate cochlear implants into new or existing rehabilitation and educational environments.

Epidemiology of Deafness

Data on the incidence, prevalence, and risk factors of deafness and hearing disorders are limited, and they do not adequately meet the needs of the clinical and basic research communities. Population-based studies are needed to determine the incidence, prevalence, and risk factors of hearing disorders, with special attention to otitis media, sudden hearing loss, age-related hearing loss, genetic epidemiological/inherited hearing loss, and noise-induced hearing loss. As part of these studies, serum and other biological samples should be obtained.

Short- and long-term research planning would be facilitated by definitive information about the scope and complexity of health problems affecting the ear and hearing. In fact, the need for well-planned and well-executed epidemiological studies pervaded Task Force discussions in all program areas, as indicated by the recommendation that the NIDCD establish a division dedicated to the epidemiology of

deafness and communication disorders. (See chapter 7, "Data Systems and Population-Based Studies.")

Identification of Congenital Hearing Impairments by Age of 9 Months

The need for early identification of congenital hearing impairment cannot be overemphasized. Every month during which a deaf infant is deprived of sound and language input aggravates the developmental consequences and adds to the difficulties of later rehabilitative management. The knowledge and methods for identifying hearing impairments during an infant's first year exist. Nevertheless, across the United States, the average age of identification remains close to 3 years. Elimination of this delay should be a top priority. The absence of data on the cost efficiency, sensitivity, and specificity of these techniques has limited their widespread use to date. Further research is needed on the evaluation of cost and effectiveness of current techniques, and continued basic research is needed to improve techniques for detection, particularly in high-risk children, and also for education of primary care physicians and parents.

Loss of Hearing During Aging

Presbyacusis, or age-related hearing loss, is the most prevalent but least studied of all handicapping conditions. Current developments in fields as diverse as physics, molecular biology, and medicine should contribute to progress in diagnosis and management. The amount of progress will depend upon a number of factors, including the application of new techniques and methods to well-designed and well-executed studies with experimental animals and human subjects. Especially critical is the application of contemporary methods from cell and molecular biology and immunology to studies of animal models of presbyacusis and to studies of the ears and brains of human subjects.

Since many factors contribute to presbyacusis, studies proposed in such related areas as homeostatic mechanisms of the ear, environmental effects on hearing and the epidemiology of hearing loss, and otopathology will contribute greatly to understanding and prevention.

Susceptibility to Environmentally Caused Deafness

The degeneration of auditory sensitivity that occurs with age even in persons not exposed to industrial noise can be considered to be the combined effect of exposure to high levels of noise associated with everyday living (sociocusis) and to ototoxic drugs, certain chemicals, and physical trauma (nosoacusis) as well as to the aging process itself (presbyacusis). All of these factors interact to an unknown degree with one's genetic predisposition. The relation between specific exposures to sociocusic and nosoacusic factors and the consequent hearing loss are as yet still largely unknown, either for a typical individual or for a specific person, because accurate data on exposure do not exist. Basic research is

needed with animal models on the mechanisms of environment-induced hearing loss and on the interaction of environmental factors on hearing. Epidemiological research focused on identification of specific individual risk factors that may lead to a greater susceptibility to hearing loss is also needed. In addition, a major program of public education is required.

Rehabilitation Devices and Behavioral Management of Hearing Loss

Rehabilitation covers the aspects of intervention that address the consequences of disordered hearing. One of two principal components is *prosthetic management*, including the fitting of hearing aids, the implantation of cochlear prostheses, and the use of tactile and other sensory aids. Related to this component is the use of other technological aids to facilitate speech training and language development. The second component is *behavioral approaches* to the prevention, circumvention, or remediation of problems arising from hearing disorders, including adaptive education for profoundly deaf and hard-of-hearing children, special training for children or adults who have difficulty with the interpretation of sound patterns (auditory agnosia), speechreading instruction, and communication therapy.

Both the prosthetic and the behavioral components rely on a thorough understanding of the characteristics of hearing disorders and of the mechanism by which their perceptual, communicative, and other consequences emerge. The following research topics are especially timely, and their results should have a major impact on rehabilitation strategies during the next few years.

Comprehensive Understanding

While much yet needs to be learned regarding the characteristics of normal hearing, it is clear that a major lack of knowledge exists in the current understanding of how perceptions are distorted with hearing loss and deafness, and of the mechanisms of the ear and central auditory system that underlie these changes. Such an understanding is basic to research on hearing aids and the development of cochlear implants and other sensory aids. That understanding is also basic to the development of truly effective programs of rehabilitation and education for the deaf and hearing impaired. Strategies, methods, and techniques for obtaining the necessary information from clinical populations are now established, and they should be applied to the hearing impaired population. This area of research should include: psychophysical studies of the distorted ear, of the aging ear, and of the electrically stimulated ear; studies of speech perception in the hearing impaired; studies of linguistic, educational, social, emotional, and vocational consequences of hearing loss; and studies of nonauditory processing disorders. Such associated disorders of the hearing impaired are often unrecognized by professionals and yet may contribute significantly to learning disabilities in the child and to emotional instability in the adult and elderly. Information in these areas may not only contribute to technical and educational

rehabilitation but also provide perspectives to enhance the understanding of basic hearing processes in the normal auditory system and of normal hearing function.

Development of Neural Prostheses

Recent advances in biomedical technology are leading to an ability to treat deafness with implantable neural prosthetic devices. Two categories of devices have evolved: one is placed directly into the cochlea and the other is placed in the auditory pathway within the central nervous system.

The peripherally placed cochlear implant is becoming an accepted method of prosthetic rehabilitation for adults who become profoundly deaf after they acquire speech and language. Improved multichannel speech processing capabilities have enhanced the performance of cochlear implants. Where early expectations were limited to hopes that patients would perceive enough sound to increase their speechreading abilities, many patients have achieved a limited understanding of spoken words without speechreading. Unfortunately, knowing who will benefit from a cochlear implant to this large extent is not yet predictable.

Even though cochlear implants have provided significant communication benefits to profoundly deaf adults, they do not restore normal hearing. Much more basic research is needed for developing the appropriate models to define the characteristics of the response of the structures of the inner ear to electrical stimulation, and also for developing neural/behavioral models to define the processing of electrically elicited activity. Future research should be directed toward improving speech coding strategies and defining the boundaries for applying this technology in the deaf population.

The advantages and consequences of using this technology with congenitally deaf children and with those who lost their hearing before acquiring speech and language remains controversial and should be the object of intensive research as well.

The safety of implantation and use of these devices can be improved. Other important topics are the development of strategies to predict cochlear implant performance before implantation and of appropriate rehabilitation strategies for patients who have received implants.

Researchers have begun developing an auditory prosthesis for placement within the central nervous system. Initial results in selected patients suggest that these patients can benefit from this approach. Future refinement of the device and development of multichannel electrodes should be pursued, using a strategy in which basic research is interfaced with technical and clinical research.

It should be emphasized that research on neural auditory prostheses is providing an additional and important window for examining auditory function, maturation, and plasticity within the central nervous system.

Development of an Implantable Hearing Aid for the Middle Ear

Conventional ear-canal-dwelling hearing aids in high-amplification applications have a tendency to display acoustic feedback and

cause discomfort, and they also have the potential to exacerbate ear infections. Compelled by the desire to overcome these problems, several research groups are pursuing the development of implantable hearing aids. If this effort is successful, the device should effectively remove the feedback problem by eliminating acoustic coupling. Such devices should provide greater comfort and benefit to hard-of-hearing people and, perhaps, be more acceptable to those who need amplification but cannot or choose not to use currently available hearing aids. This high priority area of research is likely to result in practical benefits for significant numbers of patients within a brief period of time.

Development of a Noise-Canceling Hearing Aid

Many users of hearing aids complain about the degradation of speech by environmental noise. The use of multiple microphones and digital signal processing can enhance the clarity of the speech signal in many situations. A number of studies suggest that such techniques can be developed and evaluated in the relatively near future. In the longer term, it is likely that improved understanding of impaired auditory function will permit even greater advances in this area. Many noise-reduction schemes should be applicable to other sensory aids for the deaf.

Development of Tactile Aids

It has long been recognized that the sense of touch provides an alternative channel for communicating information to the disabled. Tactile aids can provide information about the nature of environmental sounds and can provide substantial improvement in speechreading for the deaf. Both single-channel and multichannel devices have been developed. Research is needed to improve coding strategies, to match the output of tactile aids to the capabilities of the skin, and to continue the development of wearable aids. Comparisons of the effectiveness of tactile aids with neural prostheses should reveal the relative advantage of the two approaches and provide guidelines for the use of tactile aids, particularly in those cases where the patient's condition contraindicates implant procedures.

Research on Education for Hearing Impaired Children and Youth

Children with hearing impairments ranging from the profoundly deaf to the hard-of-hearing currently receive many different types of special education services. Among them are services to teach the English language to the very young hearing impaired child early enough to prevent severe educational deficits associated with severe and profound hearing impairment. These services need evaluation and improvement on the basis of rigorous data. Currently, the particular types of educational services provided vary greatly, depending on where the child lives and on the biases and training of those providing the services. Data from clinical trials should be acquired and

be made a customary part of the decision-making process for selecting education interventions for hearing impaired children.

Research should include relatively large numbers of children, studied longitudinally and randomly assigned to alternative interventions, along with compliance, blinded assessment, and analysis of cost:benefit ratios for alternative interventions.

Research on Prevention, Detection, and Diagnosis of Hearing Impairment

Trans-NIDCD issues concerning prevention and rehabilitation are discussed in chapter 7. Issues related specifically to deafness and hearing disorders are discussed below.

Children

An urgent research goal is to improve the prevention, early identification, treatment, and management of hearing disorders in young children. Activities contributing to the realization of this goal are discussed under several headings in this report, and they are summarized together below.

- Genetic factors account for more than 50 percent of severe hearing loss and deafness, both congenital and late onset. Research is needed on the genetics of deafness, specifically in areas leading to the characterization of genes involved in inner ear development and to the subsequent utilization of this information to improve diagnosis, genetic counseling, and eventual gene therapy.
- Hundreds of thousands of children destined genetically to have normal hearing are deafened by infectious agents such as cytomegalovirus, the various microbes causing meningitis, and rubella. Research is needed to develop vaccines; to understand further the organisms responsible for the disease and the ways in which they produce deafness; to describe the pathophysiological mechanisms upon which the best management strategy can be based; and to conduct clinical trials of drugs that may be used to combat these diseases.
- Research is needed on all aspects of otitis media, including studies of how it develops, how it alters the conditions necessary for normal transduction of sound, and the long-term effects of mild conductive hearing disorders on language and learning. With improved understanding of this complex disease, alternatives to current long-term antibiotic therapy or repeated surgical placement of ventilating tubes may be found, which would result in an overall savings to the health care system. There is a special need for this research on moderately and severely deaf children. The effects of even an additional mild conductive hearing loss would be much greater in a child with an existing sensorineural loss than in a normal hearing child.
- The average age at which profound hearing loss is first detected in this country is about 3 years, long after the

period crucial for language acquisition and speech development. Detection at this late stage of development has a direct negative impact on how efficiently the deaf person can adapt to life in a hearing world and/or prepare for life in the deaf community. The health care of these children is compromised by their inability to communicate adequately. Improved methods for early screening and diagnosis of hearing loss in infants and small children, especially those at risk for deafness, are needed. Similarly, current methods of screening for middle ear disease need closer scrutiny with respect to their efficiency and efficacy, and new methods should be developed if necessary.

- In most visual disorders, corrective lenses can restore normal function. The same cannot be said for sensorineural hearing loss. While amplification may overcome some aspects of the hearing loss, the total perception is often distorted. Thus, the hearing impaired child receives an abnormal auditory experience from the beginning. The results of such an experience include abnormal speech production and perception as well as learning problems. Continued emphasis is needed on developing high-technology amplification systems as well as electrical and tactile stimulation devices for those who are unable to benefit from auditory signals. Attention should also be given to habilitation strategies as a means of enhancing the overall quality of life.
- In stark contrast to the progress in other areas of research on hearing and deafness during the last three decades, methods of aural habilitation of the deaf child have changed very little. It is now appropriate to use the knowledge gained in other areas to facilitate development of more effective, data-based habilitation strategies by defining speech perception processes and by developing new technological aids for the deaf.

It is axiomatic that advances in the basic sciences lead eventually to improvements in clinical practice. Conversely, clinical research often provides insights into the normal structure and function of the auditory system. Many of the basic science studies described in this report are directly relevant to the needs of the hearing impaired child, both the deaf and the hard-of-hearing—especially studies of sensory cell regeneration, auditory development, plasticity, sensorineural transduction, and the neural coding of speech.

The Elderly

One-third of persons over 65 years of age have a hearing loss sufficient to interfere with speech perception, and the prevalence rises with increasing age. The inner ear is primarily affected, although auditory nerve and brain stem degeneration probably also contributes to the hearing disorders seen in aging persons. In most cases, no discrete cause can be found. In these cases, the term *presbyacusis* is used. Noise exposure, both occupational and nonoccupational, is often partially responsible for the inner ear damage observed. Some people are genetically predisposed to premature

loss of hearing. The role of systemic illnesses such as atherosclerosis and diabetes is poorly defined with regard to whether they are risk factors for hearing loss in aging.

The prevalence of presbyacusis has been reasonably well documented, but future population-based studies are needed to better identify risk factors, including moderate (especially nonoccupational) noise exposure, smoking, diet, and systemic disease. Such studies should also go beyond pure-tone audiometry to measure speech discrimination performance. Family studies—especially of twins—can help elucidate the genetic component. Finally, epidemiologic studies should further explore the impact of presbyacusis on affected persons and on society.

Animal models can be particularly helpful in clarifying the overlapping contributions of genetic predisposition, systemic disease, and exposure to moderate noise levels in age-related hearing loss, and can also identify different morphologic subtypes characterized by degeneration of hair cells and by structural changes in the stria vascularis (inner ear blood supply), auditory nerve, and brain. Interventions can also be studied in animal models. For example, does dietary change in animals with vascular disease reduce hearing loss while concomitantly reducing morbidity and mortality from involvement of other organ systems?

Clinical studies of patients with presbyacusis should attempt to sort the separate effects of aging and loss of sensitivity on real-world speech reception performance tests. To the extent that apparent central processing deficits are present, the deficits should be correlated with other measures of CNS structure and function derived from techniques such as MRI, PET scanning, and evoked potentials. Blood and tissue should be banked for future analyses, and collection of postmortem temporal bone and brain tissue should be a continuing area of research priority.

Clinical intervention in presbyacusis at present includes detection, measurement, and rehabilitation (including hearing aid use). As presently practiced, these methods are effective, but their positive effects on both communication skills and psychosocial well-being need better documentation. Methods of intervention can and should be considerably improved. Better hearing aids are being developed, and their efficacy and acceptability with older persons will require specific attention. Early intervention, when hearing loss is milder, may improve eventual functioning as the loss progresses. New psychoacoustic and audiologic diagnostic methods may permit identification of patients who have a poor prognosis for successful use of hearing aids, and lead to an appropriate modification of rehabilitative strategies.

Environmental Effects

Detrimental influences from the environment have long been recognized as a major contributing factor to hearing loss in the general population. A number of factors have been identified. Others have been tentatively implicated. These factors can be classified into four broad categories:

- Exposure to noise, including steady, intermittent, and impulse noise of various levels and durations. They may be

associated with occupational activities (industrial plants, airplane engines) or recreation (gunfire, rock music).

- Exposure to industrial byproducts and pollutants such as the solvent toluene and xylene.
- Administration of clinically important and potentially lifesaving therapeutics such as aminoglycoside antibiotics, polypeptide antimicrobials, diuretics, antitumor agents (cis-platin), and salicylates (commonly used as aspirin).
- Results of trauma or accidents. These may be consequences of head injuries, cranial surgery, or barotrauma.

Depending on the type and severity of the insult, the resulting hearing loss may be partial or complete, or reversible or permanently debilitating, or it may manifest itself as tinnitus. In addition, hearing loss can be exacerbated by a combination of these insults or their interaction with other factors. A genetic predisposition for individual susceptibility may also exist.

While major inroads have been made in understanding the conditions under which these agents cause hearing loss, important questions remain unresolved. Moreover, the study of many of these problems still awaits the application of modern research techniques to elucidate the molecular mechanisms leading to the impairment of auditory function. Knowledge of these mechanisms will enable the design of rational approaches to prevention, amelioration, and treatment, which are of major concern in research on these dysfunctions.

Specifically, investigations are needed into epidemiology, pathogenesis, and pathophysiology of hearing loss from environmental insults as well as into their management and prevention.

- For any suspected traumatic agent, there is the need to prove the causal relationship to the observed findings of hearing loss.
- Electrophysiological and histopathological investigations will be most helpful in determining the localization of the insult within the auditory system's anatomical substrate.
- Investigations into the molecular mechanisms triggered by each of these insults and leading up to the hearing loss are fundamental to understanding these phenomena. In addition, treatment and prevention need to be rationally based on such knowledge. These studies require the entire repertoire of modern analytical techniques of biochemistry, pharmacology, physiology, electrophysiology, and cellular and molecular biology.

In the areas of management and prevention, significant advancement can be achieved through focus on the following strategies.

- Improved methods for screening hearing loss in populations that are at risk (for example, certain industrial workers); use of high-frequency audiometry for early detection of hearing loss from pharmacologic agents.
- Investigations into the predictability of permanent hearing loss, based on temporary shifts in hearing threshold.
- Determination of safe levels for occupational noise exposure.

- Determination of the extent to which hearing impairment can be alleviated or reversed, based on early detection and removal of the insulting agents.
- Development of methods for protecting the inner ear from damage by drugs administered as pharmacologic agents. These methods, while preventing inner ear damage, must not adversely affect the efficacy of the drugs.
- Investigations into the relationship between the structure of drugs and their toxicity. This research is directed to the design of new products or the modification of existing ones so that their toxicity is attenuated while their efficacy is maintained.
- Development of procedures to test deleterious effects of drugs in models other than animals. Artificial membranes or cultures of sensory cells offer promising alternatives that need to be developed further.

This multifaceted approach, ranging from the elucidation of molecular mechanisms, through improved diagnostic and protective measures, to pharmacologic prevention and new drug design, is essential. It has the potential of providing tangible benefits to the millions of Americans now at risk of environmental harm to their hearing.

* * * * *

The foregoing describes a broad picture of the opportunities and priorities for research on hearing and its disorders. These are the headlines of a comprehensive program of research on hearing that should be supported by NIDCD during its first three years of existence. The complete plan for this research is located in Appendix E of this National Strategic Research Plan.

These recommendations are a blend of basic and clinical research, and they span a broad spectrum of the newest technologies in diverse biomedical and behavioral disciplines. They are the product of many stimulating interdisciplinary discussions among members of the Task Force and reflect their common interest in proposing a program worthy of the new national commitment to an expansion of research on deafness.

SUMMARY OF RESEARCH RECOMMENDATIONS

Major Scientific Research Opportunities

Encoding of Sound by the Inner Ear

- Analyze receptor cell activity in simple and complex sounds (including speech) and how this activity is modified through brain-controlled feedback systems.
- Elucidate the cellular and molecular mechanisms that underlie active

processes that determine the extraordinary sensitivity and exquisite selectivity of the inner ear and the basis for otacoustic emissions—that is, sounds that originate from the ear.

- Define the relationship between information in the sound signal and coded activity of the auditory nerve.
- Determine the homeostatic and immunological mechanisms (including blood flow, metabolism, and biochemical features) that maintain the normal environment of the inner ear and factors that may control regeneration and repair of sensory elements of the inner ear.

The Central Nervous System

- Define the representation and processing of signals (including speech) in the central auditory pathways, the structural and synaptic mechanisms that underlie this processing, and how these activities result in perception.
- Assess the normal life cycle of the auditory system, including the definition of the range and critical periods involved with development of auditory processes and the limits of neural plasticity in the auditory system, and define the influence of environmental and nutritional factors as well as pathological factors that compromise and distort the normal life cycle of the system.
- Encourage and support the recruitment of information and techniques from other disciplines—for instance, signal processing, artificial intelligence, molecular and cellular biology, and imaging technology.

Major Clinical Research Opportunities

Genetic Hearing Loss

- Localize and characterize the genes responsible for deafness.
- Conduct populations studies to identify the epidemiological factors of genetic deafness.

The Bases and Rehabilitation of Severe and Profound Hearing Loss

- Conduct investigations in the area of sensorineural hearing loss, in which etiological factors, the study of distortions in perceptual space, as well as the potential for sensory cell regeneration are determined.
- Continue development and assessment of the cochlear prostheses, as well as other sensorineural prostheses of the auditory system, clearly showing promise for providing speech discrimination for a significant percentage of individuals who now suffer complete isolation from the world of sound. These efforts should include studies of enhanced signal processing techniques

for the development of neural prostheses as well as hearing aids that can benefit a very large percentage of the hearing impaired.

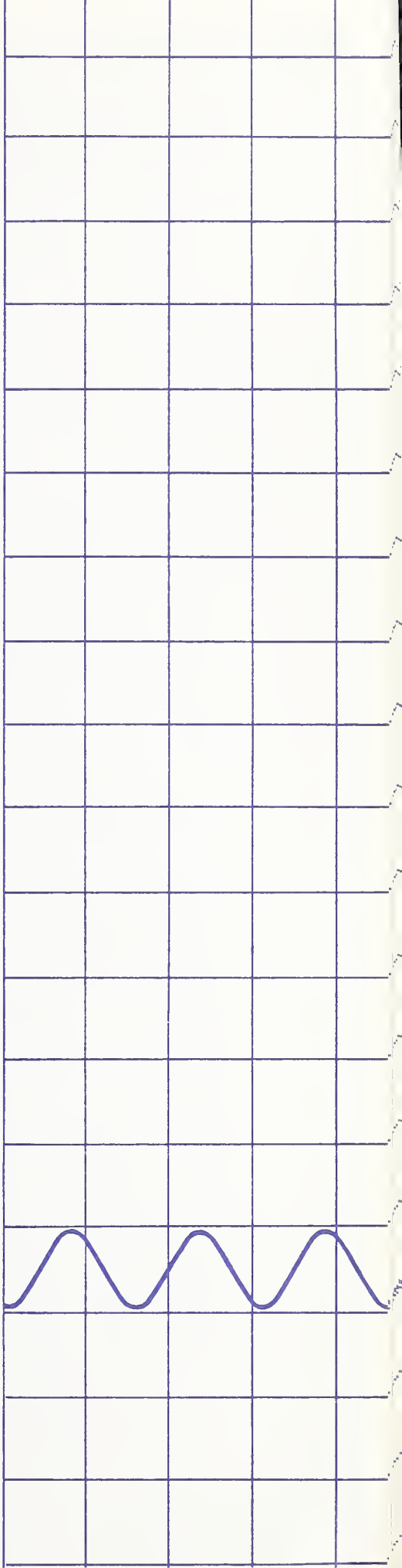
- Determine life cycle risk factors of sensorineural hearing loss, and conduct clinical trials to define the efficacy of proposed therapeutic procedures to reduce or reverse it.
- Study middle ear disease processes, including those of the inner ear bone, and the cellular and molecular mechanisms involved with otosclerosis. Also develop biochemical and immunological treatments that may inhibit this disease process, with clinical trials to assess various treatments of middle ear disease. In addition, develop vaccines to eliminate middle ear infections.
- Develop a better understanding of the basis for tinnitus and management strategies to alleviate this disorder.

Otology and Rehabilitation

- Conduct technical and behavioral studies to enhance the development of hearing aids as well as other assistive aids and their fitting for the hearing impaired.
- Define and assess the consequences of hearing disorders and the changes in perception with inner ear hearing loss, particularly in regard to the hard-of-hearing child, and the manner in which these changes may affect normal speech and language acquisition and educational development.

2.

Balance and the Vestibular System



CONTENTS

OVERVIEW 73

Symptoms of Vestibular Disorders 73
Prevalence and Cost of Vestibular Disorders 74
A Current View of Vestibular Function 75

Receptors and Reflexes 75
Integration of Information From Many Sensory Systems 77

RECENT ACCOMPLISHMENTS 78

Describing and Understanding Basic Mechanisms of Vestibular Function 78
Advances in Diagnosis of Vestibular Disorders 79
Advances in Treatment of Vestibular Disorders 80

PROGRAM GOALS 81

Understanding Basic Mechanisms of Vestibular Function 82
Clinical Studies of Vestibular Malfunction 83

Pathophysiology 83
Population Data Base for Balance Disorders 84
Diagnostic Methods and Procedures 84
Treatment Methods and Procedures 85

PROGRAM PRIORITIES AND STRATEGIES 87

SUMMARY OF RESEARCH RECOMMENDATIONS 88

Major Scientific Research Opportunities 88

Anatomy 88
Physiology 88
Chemistry 88
Psychophysics and Perception 88
Plasticity 89

Major Clinical Research Opportunities 89

Use of Pathology to Elucidate Basic Mechanisms 89
Demographics 89
Diagnostic Methods and Testing Procedures 89
Treatment Efficacy 90

2.
Balance and the Vestibular System

OVERVIEW

The vestibular system, with its primary receptors located in the inner ear, is responsible for our sense of orientation in space, for maintenance of balance and posture, and for the ability to keep objects in focus when they or we are in motion. There are five separate vestibular detectors, and their connections in the central nervous system (CNS) allow the brain to integrate information from all of the receptors with visual signals (from the eyes), cutaneous signals (from the skin), and proprioceptive signals (from muscles and joints of the body) to create what we perceive as body orientation and being at rest or in motion. It is a complex, highly interactive system capable of continual adaptations to changes in the body and in the environment. Because it has many components, there are many different symptoms that reflect its malfunction, ranging from mild discomfort to total incapacitation. Normally, balance is an automatic, unconscious process. Yet, it is essential for purposeful movement and effective communication. When balance is impaired, the capability and desire for purposeful motor activity is diminished, and there may be secondary effects on motivation, concentration, and memory.

Achieving the current view of how the vestibular system works has been a slow process, in part because the fluid-filled receptor organs that detect motion and head position are encased in bone and are hard to assess, manipulate, and study, and in part because of the complexity of the system, the many kinds of sensory information it integrates, and the multitude of symptoms that signal its malfunction.

Symptoms of Vestibular Disorders

Vestibular disorders cause a wide range of symptoms, many of which may seem unrelated to the problems of the ear but which result from the complex interactions of the different sensory modalities contributing to vestibular function and balance. The symptoms of balance disorders also vary widely, depending upon cause, location (one or both ears), the rapidity with which damage occurs, the age of the patient, and the extent of loss of function (partial or complete). Symptoms may also be intermittent, constant, or episodic. The type of symptom (dizziness versus imbalance) may also depend upon the type of movement the patient is attempting to make at the time—for instance, whether the patient is choosing a visual task, attempting to maintain an

upright stance, or both. Abnormal or unusual signals from the vestibular system, whether caused by degeneration, disease, or unusual motion environments can all result in similar or identical symptoms.

Primary symptoms arise directly from abnormal reflexes. They include: spinning vertigo, sensation of falling, imbalance, lightheadedness, disorientation, giddiness, and visual blurring (especially during movement). Secondary symptoms, which arise mostly from associated autonomic system responses to vestibular stimulation, are not directly associated with altered or abnormal orientation reflexes. They include nausea and vomiting, faintness, drowsiness, fatigue, and depression. If followed by fear, primary and secondary symptoms can lead to increased heart rate, increased blood pressure, anxiety, panic attacks, impaired memory, and decreased concentration. If severe or persistent, these symptoms will endanger the person in the work environment and result in impaired performance. Persistent symptoms also reduce the patient's willingness to participate in social functions and seriously impair the quality of life.

Prevalence and Cost of Vestibular Disorders

Balance disorders are common. Several groups of people are particularly at risk for very different reasons. The elderly suffer complications of aging, including degeneration and the inability to adapt, in addition to all the conditions that affect otherwise healthy young people. A majority of individuals over 70 years of age report problems of dizziness and imbalance, and balance-related falls account for more than one-half of the accidental deaths in the elderly. In a recent study of patients age 65 and older who have been hospitalized for disequilibrium and imbalance, the average hospital stay was four days. Furthermore, in a sample of persons age 65-75, one-third reported that dizziness and imbalance degraded the quality of their lives. Other groups of affected individuals include those with diseases of the vestibular system and healthy individuals exposed to unusual motion environments of land, water, and air and space travel.

Over 90 million Americans, age 17 and older, have experienced a dizziness or balance problem. In the United States, there are an estimated 97,000 new cases each year of Meniere's disease, a disorder that affects the inner ear and causes episodes of vertigo, fluctuating hearing loss, and tinnitus (ringing in the ear). In 1976 alone, balance problems caused over 6.7 million outpatient visits. While the majority of visits involved mild to moderate symptoms, disability was severe in an estimated 10 percent of the patients. The approximate physician charges for these visits totaled $500 million. The inclusion of cost for the associated diagnostic tests brings the full cost to several times this amount.

There are many kinds of balance disorders. Vestibular disorders may be bilateral or unilateral (asymmetric); they can result from fluctuation of function, hyperfunction, or hypofunction; or, as in the case of benign paroxysmal positional nystagmus, from mechanical distortion of the receptor organ.

Motion sickness can occur in normal individuals if the provocative stimulus is sufficiently novel, as well as in patients with abnormal vestibular function. Phylogenetically, vestibular reflexes are among the oldest known, appearing first in very primitive animals. Special environments such as diving, high-speed flying, and space travel are situations for which humans are not adapted, and genetically programmed reflexes must be overcome or reconditioned if humans are to be able to function on the frontiers of undersea, aviation, and space environments. Motion sickness, spacial disorientation in special environments, and space-motion sickness cause performance decrements that directly impact on vehicular and crew safety as well and on mission accomplishments.

A Current View of Vestibular Function

The vestibular receptor organs transduce the forces associated with head accelerations and changes in head position relative to gravity. The result is excitation of nerves going to brain control centers, which use these signals to develop a sense of orientation and to activate muscles that subserve automatic movements of the eyes, locomotion, and posture.

Many of the vestibular pathways in the central nervous system are organized into reflex pathways that serve to stabilize and coordinate movements of the eyes, head, and body. These processes are collectively referred to as the vestibular reflex systems.

While the basic receptors and neural structures are genetically determined, the development and fine-tuning of vestibular function is a dynamic process that is dependent upon use and upon interaction with the environment during early development and throughout life. There must be an ongoing process of calibrating reflexes in order to adapt to physical changes in the musculoskeletal system to compensate for the pathologies (for example, inner ear disease or visual losses), and to adapt to changes in the environment, such as an altered gravitational field, as in space. The proper responses to vestibular information require learning and memory.

Receptors and Reflexes

Two of the vestibular end organs, the saccular and utricular maculae, sense linear forces acting on the head. The cristae of the three semicircular canals monitor rotatory acceleration forces. Transduction involves excitation of specialized hair cells located in the walls of the maculae and semicircular canal cristae. Hair cells contain ciliary bundles ("hairs") protruding from their apical (top) surfaces. The cilia are embedded in gelatinous structures—the cupulae of the semicircular canals and the otoconia membranes of the maculae—that act to couple the forces acting on the head and result in the bending of the cilia. Displacement of the cilia regulates the opening of the transduction channels in an electrical response of the hair cells. The electric response controls the release of an excitatory nerve transmitter at the hair cell-afferent

synapse, which results in coded nerve impulses. Even at rest, hair cells continue to release neurotransmitter with the result that afferent nerve fibers have a background level of activity. Bending the cilia in one direction causes an increase in transmitter release and nerve activity; bending in the other direction causes a decrease. The receptor organs are arranged in the temporal bone in a spatial arrangement, so as to detect linear and angular accelerations in three dimensions.

The hair cells are innervated by afferent vestibular nerve fibers, which transmit nerve impulses to the vestibular nuclei, brain stem, and cerebellum. These structures then project to brain motor centers controlling the eye, head, and body muscles. The vestibular receptors are also provided with an efferent innervation, which originates in the brain stem and serves to modulate the activity and function of the receptors. Furthermore, the organization of signals from the two ears is complementary, so that head forces that lead to the excitation of a sensory organ on one side also cause inhibition of the corresponding end organ on the opposite side. For example, with head rotation to the right, there is a proportional increase in the firing of afferents in the right horizontal semicircular canal nerve and a decrease in the corresponding nerve on the left side. Hence, the activities of the left and right ear semicircular canals are reciprocally organized. In addition, some of the neurons in the vestibular nuclei on one side are excited, while others are inhibited. By means of these two classes of neurons and the reciprocal activation of the receptors in the two ears, groups of muscles are reciprocally activated, so that movement can occur in a push-pull manner.

One such reflex, the vestibulo-ocular reflex (VOR), causes the eyes to move in reaction to head motion. When the head is rotated to the right, the horizontal semicircular canal is excited in the right ear and is inhibited in the left ear. By way of reciprocal reflex pathways projecting to the oculomotor nuclei (neurons that innervate the muscles of the eyeball), the eyes move to the left by the same angle as the head movement. As a result, the eyes are still directed to the original point in space. Thus, gaze is stabilized, and clear vision is maintained.

Despite its seeming simplicity, the operation of the VOR raises important questions about vestibular function. The effective stimulus to the canals is the force associated with head angular acceleration. Because of viscous drag, the angular deviation of the cupulae and the associated afferent nerve response are appropriately proportional to the velocity of the head rotation. But the required eye movement is related to the change in head position, not to head velocity. How does the central nervous system transform an afferent nerve signal related to head velocity into a motor command that can produce an eye movement that compensates for the change in head position? The answer to this question requires the solution of several other problems. What are the chemicals released by stimulation of hair cells, and how do they control afferent nerve impulses? How do different types of hair cells and nerve fibers contribute to the encoding of head motion? How are the dynamics of the afferent signals transformed

in central pathways? How do different central neural structures contribute to this and other reflexes? What are the precise pathways that convey such finely tuned coordination of motor and postural reflexes? How do the reflexes remain calibrated in the face of alterations in their normal organization and function? Work done over the past decade has provided tentative answers to several of these questions, but much remains to be solved.

Integration of Information From Many Sensory Systems

Orientation is a complex function that allows persons to know the relationship of their body parts to the environment in a dynamic and reciprocal interaction. Balance is the process that allows persons to move or maintain their body and body parts in a specific relationship to the environment. Maintaining orientation and balance requires the integration of information from many sources (especially the ears, eyes, skin, muscles, and joints) and the transformation of these signals into coordinated patterns of muscle activity. To maintain balance effectively under a variety of sensory and support surface conditions, the combinations of sensory signals and muscle activity patterns must be interpreted appropriately. Sometimes information from two or more systems disagree, and the brain's higher centers must quickly decide which information is correct. An example of such a sensory conflict situation arises when the driver of an automobile is waiting at a traffic light and an adjacent car begins to move forward. The driver initially senses that he is drifting back and presses the brake, only to realize that it is his visual surround and not he, himself, that is moving. Another example: when one stops spinning, as when one gets off a merry-go-round, the world still seems to move.

The inertial-gravitational reference provided by the vestibular system is critical to the resolution of sensory conflicts between visual and vestibular and between spinal and vestibular inputs. The vestibular inputs are critical to the selection of appropriate postural movement strategies. The cerebellum and basal ganglia are important central neural structures that help to mediate visual-vestibular-proprioceptive interactions and coordinate proprioceptive reflexes subserving balance. While it is clear that the higher cortical centers are also important, their precise role remains to be determined.

Because vestibular signals interact with all of the major sensory systems and involve major brain centers, a large number of disease processes can impair balance; and because balance mechanisms operate at an automatic and unconscious level, patients frequently cannot adequately identify or describe their balance problems. The physician often finds it equally difficult to distinguish among the many possible causes for imbalance and dizziness.

Motion sickness can occur with exposure to conflicting orientation cues, perceptions of space, and motion signals. Although motion sickness is one of the most common causes of vestibular symptoms, the mechanisms underlying its production, its clinical manifestations, its palliation, and its

adaptation to motion are not understood. Motion sickness occurs both in normal people who are exposed to unusual motion conditions and in patients with vestibular disorders. A very high incidence of motion sickness occurs in pilots being trained in stationary flight simulators (*New York Times* 2/20/89, p. D1). Trainees may learn behavioral responses on such simulators to prevent development of motion sickness, which would be inappropriate in a real aircraft in flight and thus could endanger themselves, their passengers, their missions, and their aircraft. Similarly, space motion sickness, which affects one-half of astronauts, is a major unsolved problem for space travel.

The current understanding of vestibular function has allowed the application of computer-based measurement technology to detect abnormalities in the movements of eye and postural control muscles as an aid to the diagnosis of underlying disorders.

RECENT ACCOMPLISHMENTS

Describing and Understanding Basic Mechanisms of Vestibular Function

During the past decade, significant creative research has been conducted on the anatomy and physiology of the vestibular end organs, balance and gaze mechanisms, and central neural pathways. Ideas and concepts that have been explored in both basic science and clinical laboratories have led to the development of new protocols and computer-based techniques, which can help in the evaluation of gaze and balance disorders. There are now many laboratories in the United States where this technology is used to make precise, quantitative measurements of vestibular reflexes, posture, and the interactions of different sensory inputs to the central nervous system.

Significant accomplishments include the following:

- Some important biophysical properties of hair cells have been defined, including a characterization of transduction mechanisms, which show how the hair cells of the inner ear detect forces exerted upon them that result from motion of the head, and transform this activity into nerve impulses.
- At the light and electron microscopic level, the innervation patterns of the vestibular end organs and the relationship between anatomical characteristics and changes in the discharge of afferent nerve fibers due to physiological stimulation have been partially elucidated.
- Some information has been obtained about the responses of single vestibular afferent fibers to electrical stimulation of efferent neurons in the brain stem.
- The use of behaviorally conditioned preparations and new techniques for localizing and labeling single neurons has led to a better understanding of the interaction of different vestibular and visual inputs. The complexity of the

pathways involved in the interaction of reflex responses is being elucidated.

- Preliminary data have been gathered on projection patterns from the vestibular end organs onto the vestibular nuclei in several animal species.
- There is better understanding of the convergence of signals carrying information derived from other sensory modalities such as vision and proprioception (due to information from muscle sensors) onto single neurons within the vestibular nuclei and the cerebellum.
- The dynamic characteristics of signals generated by single neurons in the vestibular nuclei and other brain stem structures are defined under a variety of conditions.

Perhaps the most remarkable property of the vestibular and balance control systems is their ability to maintain useful functioning response to many novel motion environments and to adapt to abnormal function in one or more of their components. The ability to systematically adapt sensory, neural, and motor components of these systems in order to achieve useful compensation must be more thoroughly understood. Before advances in vestibular health care, particularly rehabilitation, can be developed from sound scientific concepts, the significance of CNS phenomena (such as neuronal sprouting, reactive synaptogenesis, and deafferentation supersensitivity), which are thought to play important roles in adaptive control, must be correlated systematically with behavior. Some accomplishments that have opened exciting new areas of investigation include:

- A recognition of the essential role of adaptive plasticity in the normal function of the vestibular system. The vestibular system is unusual in that it is not static. It is constantly changing in response to changes in the environment or internal elements (for example, inner ear pathology or CNS diseases).
- Identification of some features of the neural pathways that underlie this constant change in animals.
- Development of more sophisticated mathematical models of vestibular reflexes (for example, the VOR), which include multisensory inputs, three-dimensional spatial coordinate systems, gaze velocity storage, and internal feedback (that is, efferent copies or transformations of command signals driving the motoneurons).
- Acquisition of some preliminary information about transmitter properties of neurons in the vestibular end organ and the central nervous system in some animal models.
- An anatomical description of the basic circuitry in the brain stem subserving the VOR.

Advances in Diagnosis of Vestibular Disorders

Provision of rational therapy for vestibular disorders requires a knowledge of what parts of the vestibular system cause different symptoms or syndromes. The

knowledge of such correlations is still very primitive. In the past decade, some progress has been made in the effort to identify these relationships.

Several kinds of vestibular and balance disorders can be recognized. Peripheral vestibular disorders may affect one or both ears, and such disorders can occur suddenly or gradually. They may result in an increased or decreased neural activity, but one important source of malfunction is a disruption in the normal balanced flow of information from the two ears. Because the system is normally so adaptive, some gradual changes and loss of function may not even be apparent to the patient.

Newly available computer-based technology has enabled quantification of both VOR and vestibulo-spinal reflex (VSR) abnormalities. For the VOR, new methodology that employs computer-controlled rotation chairs and eye movement analysis has enabled quantitative measurement of both caloric and rotational eye movement responses in the clinic. More powerful rotation devices have permitted the development of visual-vestibular interaction (VOR) tests at more physiologic frequencies (0.5 Hz or above) in association with controlled movements of visual surrounds in order to evaluate compensatory mechanisms. Computer-controlled visual surrounds and support surface around a standing human (moving platform posturography) can separate inner ear, visual, and muscle stretch contributions to balance. Both the VOR and VSR techniques identify, quantify, and localize the source of vestibular and balance disorders.

Magnetic resonance imaging (MRI), with or without the addition of intravenous contrast material, has provided a recent major advance in the ability to diagnose a variety of disorders that affect balance. Among these are space-occupying lesions (for example, eighth nerve and other tumors, aneurysms and cysts affecting the brain, cranial nerves, and the inner ear structures) and several degenerative brain disorders (for example, vascular lesions, multiple sclerosis, and ataxia syndromes). Dynamic computerized tomography (CT) with contrast permits the evaluation of blood flow in various parts of the brain stem and brain. Digital subtraction angiography and Doppler flow studies have facilitated screening for cerebrovascular disease, including arteriosclerosis, relating to balance disorders. Positron emission tomography (PET) is currently under development for use in the study of central vestibular pathways.

Advances in Treatment of Vestibular Disorders

Diagnostic methods now allow the identification of many disorders of the vestibular system that are amenable to effective medical management or rehabilitation (for example, various types of endolymphatic hydrops, vestibular neuronitis, vascular deficiencies, ototoxicity) as well as those that require surgery. Surgical therapy of vestibular disturbances has advanced in some areas during the past decade through the development of new or the modification of old techniques. Successful repair of fluid leaks from the inner ear to the middle ear air spaces is effective in controlling

dizziness caused by rupture of these structures. The efficacy of surgical management for vestibular disorders has not been established for most procedures now in use. Prospective randomized clinical trials need to be conducted to validate these procedures.

Thanks largely to improved diagnostic tests, smaller acoustic neuromas and other tumors can now be discovered, often while hearing is still serviceable. This advance has led to earlier surgery, with a reduced risk to the patient, particularly if the tumor is discovered before compression of the brain stem occurs. Facial nerve and auditory monitoring, as well as improved anesthesia techniques, add to the safety of these operations and reduce the incidence of complications such as facial palsy. A variety of lasers and the ultrasonic aspirator facilitate tumor removal.

Operations designed to interrupt the pathways from the inner ear to the brain have also gradually evolved. The singular neurectomy operation (section of the posterior semicircular canal nerve, also known as the singular nerve) arose from an understanding of the pathological mechanism in benign paroxysmal positional nystagmus and vertigo. The selective vestibular nerve section with preservation of hearing evolved from the earlier practice of severing the entire eighth cranial nerve. Vestibular nerve section has replaced labyrinthectomy (surgical excision of the entire inner ear) for incapacitating vertigo in patients with usable hearing in the diseased ear. Better understanding of the pathophysiology of vestibular deprivation has led to more rational medical treatment of patients with various disorders (such as vestibular neuronitis, labyrinthitis, and developmental and degenerative disorders of balance). The vestibular role in various disorders of imbalance due to both inner ear and CNS lesions (disequilibrium, ataxia, unsteadiness, dizziness) is being better understood with research advances.

Many vestibular disorders due to metabolic abnormalities or associated with psychogenic factors (for example, panic attacks) can now be adequately managed. Some autoimmune disorders and ataxia syndromes may now be diagnosed and treated appropriately with medication.

PROGRAM GOALS

There is a need to expand the knowledge of normal and abnormal structure and function of the vestibular system using modern anatomical, electrophysiological, immunological, histochemical, genetic, molecular biology, and other recently developed techniques. Scientists must be trained and support mechanisms developed to sustain this research effort long enough to establish a knowledge base commensurate with other fields of medicine. Both human pathological material and animal models should be used. Most of the goals identified for balance system disorders in the 1979 *Report of the Panel on Communicative Disorders* (NIH Publ. No. 79-1914, pp. 298-316) have not yet been accomplished.

Understanding Basic Mechanisms of Vestibular Function

Because there is an integration of the information from several different sensory modalities, and because the sensory-to-motor transformations involve the activation of many different muscle groups, there are special challenges in studies of the neural mechanisms underlying the control of balance and orientation. A systematic investigation should include studies at different levels of both the peripheral and central nervous systems.

At the level of vestibular inputs to the central nervous system, detailed studies of the vestibular receptors are needed, such as a definition of the ultrastructural, biophysical, and biochemical properties of the various components that participate in the transduction process. The following goals have been identified:

- Characterize the biochemical properties of vestibular hair cells, including transmitter chemistry.
- Elucidate the biophysical properties of the hair cells and specializations within different regions of the end organs.
- Define the role of efferent neurons, including an analysis of the effects of efferent activation on the receptors and afferent synapses. The process of afferent-efferent interaction needs to be defined in terms of electrical, chemical, metabolic, and neuromodulatory phenomena.
- Characterize the mechanisms for regulating fluids, electrolytes, and other metabolic compounds in the inner ear.
- Define the action of pharmacologic agents on end organ structures.
- Develop experimental models for peripheral vestibular dysfunction, and take advantage of species differences and genetic variations.

At the level of the central nervous system, there is a need for a physiological and anatomical description of the neurons responding to stimulation of the different vestibular end organs (both canals and otoliths) and other sensory inputs (visual and muscle proprioceptive), an anatomical and functional description of the neural connections within the vestibular nuclei, brain stem, and cerebellum, and projections from these areas to motoneurons and to neurons in other higher centers responsible for visual and spatial stabilization. The following goals have been identified:

- Correlate the morphological characteristics with the neurophysiological characteristics of neurons within different parts of the vestibular nuclei during isolated and combined canal, otolith, and visual stimulation.
- Define the convergence patterns of different vestibular receptors onto the vestibular nuclei neurons.
- Define the convergence patterns of different sensory systems (visual, proprioceptive) and other brain structures (reticular formation, cerebellum, and higher motor and sensory centers) onto the vestibular nuclei neurons.
- Determine the transmitters and modulators for the vestibular neurons and projections to the vestibular nuclei.

At the behavioral level, there is a need to define the strategies that the central nervous system uses to control movement and posture and a need to define the relative but different roles played by the sensory inputs from the semicircular canals, otolith organs, neck muscle proprioceptors, and the eyes to the dynamic characteristics of movement. Also, there is a need to elucidate the mechanisms by which vestibular inputs contribute to the perception of the earth vertical and body position in space (in an inertial reference frame) and the factors that confound that perception.

The adaptive plasticity of gaze has been recognized, but the neural mechanisms are not understood. In light of the demands placed on the central nervous system for maintaining balance during normal growth and development as well as during exposure to unusual environmental conditions such as altered gravitational fields, there must also be significant adaptive changes for the balance mechanisms. The mechanisms of the central nervous system for maintaining balance have not been systematically studied. The following goals have been identified:

- Define the anatomical, physiological, and biochemical changes in the neural circuitry that underlie adaptive plasticity.
- Develop models for systematic investigation of how adaptation contributes to the compensation for deficits in the vestibular system.
- Investigate psychophysical aspects of normal vestibular function, and develop standards for evaluation of normal vestibular sensations.
- Develop an understanding of the types of physical stimuli that govern adaptive behavior, as the relative importance of the various inputs (inputs from the foveal versus retinal periphery, inputs from visual versus vestibular end organs, inputs created by active versus passive motion) are yet to be defined.
- Develop an understanding of the dynamic neurochemical bases of the adaptive processes, since these mechanisms are most important for understanding everyday orientation and its recovery following pathological changes.
- Conduct psychophysical studies to better understand the imprinting of vestibular experiences in the development of psychological profiles in normal subjects and patients.

The insights provided by information about pathological states should increase the understanding of normal function and possible protective or rehabilitative strategies.

Clinical Studies of Vestibular Malfunction

Pathophysiology

There is a dearth of knowledge about the etiology, pathophysiology, diagnosis, management, prevention, and rehabilitation of balance disturbances. Notable among these are Meniere's syndrome, benign paroxysmal positional vertigo, vestibular neuronitis, drug and environmental ototoxicity, terrestrial and space motion sickness,

labyrinthitis, and central balance disorders including cerebrovascular, neoplastic, congenital, aging, developmental, and degenerative changes, and trauma, infectious disease, and allergic disorders. These disorders lend themselves to a broad-based multidisciplinary approach that should be addressed by the National Research and Training Centers (NRTCs) for Deafness and Other Communication Disorders. (See chapter 7.) These studies should include shared data bases, research training, and basic clinical research components, including psychosocial components, nursing, and rehabilitation.

Population Data Base for Balance Disorders

There is an urgent need for a population data base to determine the prevalence of vestibular disorders and balance dysfunction. The prevalence of various etiologies; the signs, symptoms, and risk factors associated with these etiologies; the outcome of diagnostic procedures; and the efficacy of treatment should be recorded.

Since individual centers are unlikely to generate sufficient numbers of patients in certain categories of disorders, an appropriate data base should be developed to receive data from medical centers across the country that treat a large number of dizzy patients. There is a need for a multidisciplinary advisory group. Research goals include the following:

- Study the basic mechanisms of compensation of injury to all components of the balance system.
- Develop cooperative studies using resources of the National Institutes of Health, the Uniformed Services University of the Health Sciences, and the National Aeronautics and Space Administration to investigate balance problems in special environments, such as under water, in high-performance aircraft, and in space. Included are conditions of barotrauma involving intralabyrinthine rupture and hemorrhage, and decompression sickness.
- Develop normal data on temporal bones and CNS tissues in the same individuals. Normal data are needed across all age ranges, from early years (developmental) and late years (aging). Before death, these individuals should be carefully evaluated. Data should include history, physical examination, and modern tests of auditory and vestibular function and central imaging (CT or MRI).

Diagnostic Methods and Procedures

- Determine the sensitivity, specificity, and clinical efficiency of all present and future diagnostic methods for all major categories and classifications of vestibular abnormalities.
- Encourage and facilitate development of new diagnostic methods for vestibular and other balance disorders. New tests should be able to localize (that is: left ear, right ear, or both ears; brain stem; or neuromuscular or autonomic nervous

system) as well as quantitatively describe altered systems function and dynamics. Feasibility studies should evaluate vestibular-evoked responses and vestibular electrical stimulation as diagnostic procedures for balance disorders. These techniques should be developed for all age groups.

- Develop diagnostic procedures based on measurements of interrelated auditory and vestibular systems, such as electrocochleography and middle ear biophysical measures.
- Apply basic science concepts and clinical tests to understand the development of vestibular function in normal individuals and patients with congenital or birth defects.
- Develop computer simulation of sensations associated with balance disorders in order to establish objective measures of symptoms.
- Develop functional tests of otolith and vertical canal interactions.
- Develop better psychophysical methods for assessing sensation of motion.
- Use existing technologies such as MRI, PET scanning, and magnetoencephalometry to assess abnormalities that are difficult to diagnose, such as hydrops or isolated brain stem dysfunction.
- Develop disease-specific immunological techniques for inner ear, autoimmune, and tumor diagnosis.
- Develop cross-sectional and longitudinal data bases of sensorineural balance system information concerning: (a) temporal bone histopathology, (b) the central and peripheral balance systems, and (c) standardized balance function tests.
- Establish a good description of normal postural development in children.
- Identify factors that affect vestibular development.
- Further develop genetic models in animals of human vestibular deficiencies. Of the five mutations known to affect the development of vestibular otoconia in inbred species of mice (the calcium carbonate crystals in the inner ear organs, which stimulate hair cells in response to gravity and motion), three also affect pigmentation. Their expression can be prevented by dietary supplementation during key periods of fetal development.
- Investigate mutants with auditory anomalies to determine possible associated vestibular anomaly.
- Develop quantitative measures of vestibular function in infants and children.
- Develop genetic mapping techniques for congenital and neoplastic balance system disorders. Apply recently developed molecular biology techniques to the study of the pathology and pathophysiology of balance disorders, with special emphasis on disorders of unknown etiology.

Treatment Methods and Procedures

Current treatment methods are constrained by a lack of understanding of underlying disease processes and mechanisms.

Medical treatment of balance disorders arising from the inner ear often consists of prescribing medicines that suppress vestibular activity, which often have troublesome side effects.

Current surgical treatments are usually based upon destructive concepts. Notable exceptions are repair of perilymph fistulae, chronic ear infections (cholesteatoma), and endolymphatic sac procedures. Consequently, methods of both medical and surgical treatment need to undergo scientifically rigorous evaluation, including outcome analysis followed by cooperative or multi-institutional clinical trials.

Cooperative studies that could be centrally administered and supported by the NIDCD and other agencies and organizations should be encouraged as part of the recommended NRTCs (see chapter 7).

A more focused program for research on treatment methods includes:

- Evaluation of therapies that are safe and effective in a variety of settings, including hazardous work environments.
- Pharmacological methods based upon a thorough understanding of the basic neurotransmitter systems involved in balance.
- Rehabilitation research to aid those who have suffered disorders of balance function as an adjunct to medical or surgical treatment.
- Further evaluation of balance training and movement strategy exercises for possible use in at-risk populations. These exercises may also have a use in preventive medicine, rehabilitation, and treatment of motor impairments associated with learning disorders.
- Development of appropriate animal models for all major classes of balance disorders.
- Identification of balance system toxic agents in the occupational environment and in therapeutic agents.
- Development of restorative surgical procedures, including prosthetics, for treatment and rehabilitation of balance disorders.
- Development and evaluation of assistive devices for balance disorders.
- Investigation of hormonally mediated phenomena of morning sickness associated with pregnancy, the exacerbation of endolymph hydrops premenstrually, and their similarity to motion sickness.
- Investigation of the ubiquitous psychological epiphenomenon associated with balance disorders.
- Evaluation of the patient-reported relationship of vestibular symptoms, including objective balance abnormalities, with panic disorders.

The Task Force stresses both the enormity of health care problems related to vestibular disorders and the paucity of research efforts devoted to their solutions. The many needs include basic and clinical research training, with immediate emphasis upon the development of leadership needed to spearhead and sustain basic and clinical research programs. NIH efforts should be actively coordinated with other agencies and organizations with interests or needs in vestibular and balance disorders including

the National Aeronautic and Space Administration, the Armed Forces, the National Science Foundation, universities, foundations, institutes, and industry. There is an urgent need for interdisciplinary research efforts utilizing both basic and clinical research teams. Centers of excellence should focus upon the rigorous application of research principles to the study of the pathophysiology and management of patients with clinical balance disorders. These centers should include in their protocol plans for training and development of young basic and clinical investigators.

PROGRAM PRIORITIES AND STRATEGIES

There are special problems in developing a level of understanding of vestibular and balance disorders sufficient to allow appropriate diagnosis and treatment. A large portion of the needed information can be obtained only by clinical research, which requires the active participation of researchers with expertise in several medical disciplines (including otolaryngology, neurology, ophthalmology, and pathology) collaborating with basic and clinical scientists with expertise in several fields. Similarly, the information needed to answer basic science questions must come from several distinct disciplines. Obtaining it will require the coordinated efforts of investigators trained in anatomy, chemistry, physics, and molecular biology.

Since there is already a critical shortage of clinician investigators and basic scientists suitably prepared to undertake the work, a high priority must be to increase research personnel resources. Several innovative methods for accomplishing this objective are outlined in chapter 7, "Research Training and Research Personnel Resources."

The recommendation for the creation of NRTCs is critical, especially for vestibular studies. Centers facilitate multidisciplinary studies, encourage the recruitment of physicians and scientists to vestibular research by placing them in training environments where the necessary support facilities and instrumentation are available, and encourage basic scientists to engage in vestibular research in an atmosphere of clinical coordination and support.

Another great need in the area of vestibular and balance disorders is an upgrading in the level of training of primary medical care physicians and relevant professionals in the recognition of balance disorders. They should understand the need for appropriate referral for special testing and the availability of appropriate treatments and habilitative or rehabilitative procedures. Education of professionals can be facilitated both within the framework of training centers and through the use of an information clearinghouse.

There is also a great need for a centralized, well-publicized source of information about centers for diagnosis and treatment, and national, state, and local support groups for sufferers of various manifestations of balance disorders and for their families. An information clearinghouse would provide such a service and support

the efforts of the groups presently struggling to meet this need. (See chapter 7.)

SUMMARY OF RESEARCH RECOMMENDATIONS

Major Scientific Research Opportunities

Anatomy

- Elucidate the microscopic anatomy of receptors (structure, innervation patterns, molecular biology).
- Correlate the morphological characteristics with the neurophysiological characteristics of neurons within different parts of the vestibular nuclei during isolated and combined canal, otolith, and visual stimulation.
- Define the distribution and convergence patterns of different vestibular receptors onto the vestibular nuclei neurons, both anatomically and functionally.
- Define the convergence patterns of pathways conveying information from different sensory systems (visual, proprioceptive) and other brain structures (reticular formation, cerebellum, and higher motor and sensory centers) onto the vestibular nuclei neurons.

Physiology

- Elucidate the biophysical properties of the hair cells and specializations within different regions of the canal and otolith end organs.
- Develop an understanding of the types of physical stimuli that govern adaptive behavior, such as the relative importance of different inputs (for example, inputs from the foveal versus retinal periphery, inputs from visual system versus vestibular end organs, and inputs created by active versus passive motion).
- Develop experimental models for peripheral vestibular dysfunction, and take advantage of species differences and genetic variations.

Chemistry

- Characterize the biochemical properties of vestibular hair cells, including transmitter chemistry.
- Define the action of pharmacologic agents on end organ structure.
- Determine the transmitters and modulators for the vestibular neurons, projections to the vestibular nuclei, and projections to and among higher central nervous system nuclei subserving balance and orientation.

Psychophysics and Perception

- Investigate psychophysical aspects of normal vestibular function, and develop

standards for evaluation of normal vestibular sensations.

- Conduct psychophysical studies to understand better the imprinting of vestibular experiences in the development of psychological profiles in normal subjects and patients.
- Evaluate the patient-reported relationship between vestibular symptoms, objective balance abnormalities, and panic disorders.

Plasticity

- Define the anatomical, physiological, and biochemical changes in the neural circuitry that underlie adaptive plasticity.
- Define the basic mechanisms of compensation for injury to different components of the balance system.
- Investigate balance problems in special environments, such as under water, in high-performance aircraft, and in space.

Major Clinical Research Opportunities

Use of Pathology to Elucidate Basic Mechanisms

- Further develop genetic models in animals of human vestibular dysfunction.
- Investigate mutants with auditory anomalies to determine possible associated vestibular anomalies.
- Develop genetic mapping techniques for congenital and neoplastic balance system disorders, using recently developed molecular biology techniques to study the pathology and pathophysiology of balance disorders, with special emphasis on disorders of unknown etiology.

Demographics

- Establish normative histological and immunocytochemical data on temporal bones and central vestibular nervous system tissues in the same individuals.
- Establish a quantitative profile of normal vestibular reflexes, including postural development in children.
- Establish data banks to assess the distribution of vestibular disorders among different sectors of the population.
- Identify toxic agents in the occupational environment and in therapeutic agents that impair the balance system.

Diagnostic Methods and Testing Procedures

- Determine the sensitivity, specificity, and clinical efficiency of all present and future diagnostic methods for all major categories and classifications of vestibular abnormalities.

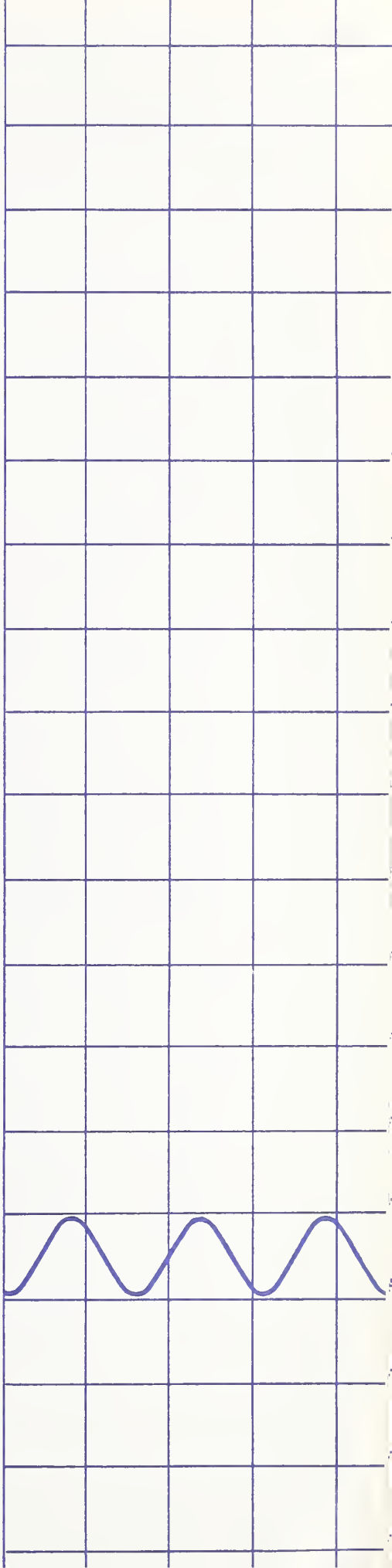
- Encourage and facilitate the development of new diagnostic methods and procedures for documentation of vestibular and other balance disorders.
- Develop diagnostic procedures based on measurements of interrelated auditory and vestibular systems, such as electrocochleography and middle ear biophysical measures.
- Develop a model for computer simulation of sensations associated with balance disorders in order to establish objective measures of symptoms.
- Develop functional tests of otolith organ, vertical semicircular canal interactions, and interactions of these sensory inputs in the central nervous system.

Treatment Efficacy

- Evaluate pharmacologic therapies to insure that they are safe and effective in a variety of settings, including hazardous work environments.
- Establish outcome analysis methods for the medical and surgical management of major disorders of the vestibular and balance systems.
- Evaluate balance training and movement strategy exercises for possible use in at-risk populations.
- Develop and evaluate assistive devices for balance disorders.

3.

Voice and Voice Disorders



CONTENTS

OVERVIEW 97

Emergence of Voice Science and the Science of Voice Disorders 97
Definition of the Voice and Speech Systems and Their Disorders 97
High Priority Areas of Investigation 98

Long-Term Priority Areas 98
Short-Term Priority Areas 98

Background 99

RECENT ADVANCES 100

Photography of the Larynx 100
Acoustic Measurements 101
Measures of Vocal Fold Vibration 101
Imaging the Larynx and Pharynx 102
The Neurophysiology of Laryngeal Movement 102
Advances in the Diagnosis and Treatment of Voice Disorders 103

Vocal Fold Neoplasms 103
Vocal Fold Paralysis 103
Disorders of Vocal Fold Movement 104

RESEARCH STRATEGIES AND PRIORITIES 104

Normal Structure and Function 104

Laryngeal and Respiratory Physiology 104
Biomechanics of the Larynx 105
Cellular and Molecular Biology and Anatomy 105
Regulation of Pitch, Loudness, Register, and Vocal Quality 105
Perturbation Analysis 106
Source-Tract Interactions 106
Vocal Fatigue 107
Role of the Larynx in Swallowing 107
Neural Regeneration 107

Development 107

Validation of Animal Models 107
Embryology 108

Comparative Anatomy 108
Critical Periods 108
Vocal Control 109
Effects of Deafness on Developmental Vocal Control 109

Aging 109

Cell Death 109
Anatomical, Cellular, and Molecular Biology 110
Muscle Atrophy 110
Deterioration of Joints, Ligaments, Membranes, and Other Tissues of the Laryngeal, Pulmonary, and Secretory Systems 110

Exceptional (Trained) Vocal Behavior 111
Diseases and Disorders 111

Structural 111

Congenital and Acquired Laryngeal Pathology 111
Inflammatory and Infectious Diseases of the Upper Airway and Larynx . . . 112
Tumors of the Upper Aerodigestive Tract 112

Neural 113

Vocal Fold Paralysis 113
Spasmodic (Spastic) Dysphonia 113
Laryngeal Disorders Secondary to Neurological Disorders 114

Idiopathic Dysphonia 114
Environmental Factors 114
Recovery From Surgery and Injury 115
Endocrine and Pharmacological Voice Changes 115
Respiratory Tract Disorders 115
Disorders Secondary to Other Disorders 116
Dysphagia 116

Techniques, Procedures, and Devices for Analysis and Treatment 116

Diagnostic Aids 116

Electromyography 117
Acoustic Analysis 117
Aerodynamic Measures 117
Imaging 118

Prostheses 118
Surgical Procedures 119
Medical and Behavioral Treatment 119

Training Needs in Voice and Voice Disorders 121
Research and Training Centers 122
Epidemiology 122

SUMMARY OF RESEARCH RECOMMENDATIONS 122

Major Scientific Research Opportunities 122

Laryngeal and Respiratory Physiology 122
Biomechanics of the Larynx 123
Cellular and Molecular Biology and Anatomy 123
Development, Critical Periods, and Aging 123

Major Clinical Research Opportunities 124

Organic, Idiopathic, and Neurologic Diseases 124
Procedures for Diagnosis and Treatment 125
Instrumentation and Technological Developments for Diagnosis and Treatment . 126

3.
Voice and Voice Disorders

OVERVIEW

Emergence of Voice Science and the Science of Voice Disorders

Vocalizations such as laughing, crying, moaning, singing, and shouting can exist outside the speech context. Infants, for instance, produce a variety of vocalizations, as do many animals. As human speech develops, modulations of the basic source of sound combined with additional sources create a large inventory of speech sounds.

Certain disorders of voice occur, and some can be distinguished from speech disorders. Many people who have acquired normal speaking skills become communicatively impaired when the vocal apparatus fails. This impairment is significant in modern society, particularly in noisy and polluted environments, where much demand is placed on speech and oral communication. In addition, there are increasingly larger aged and hearing impaired populations, and populations under psychological and physical stress. Since voice production is not the primary function of the larynx, heavy demands on this organ, as well as its vulnerability to infectious diseases, require special attention.

Within the last decade, the field of voice science and the clinical specialties of phonosurgery and voice therapy have expanded in several ways. A large new knowledge base has been developed, and a clinical delivery system has emerged resulting in recognition, diagnosis, and treatment of patients with voice impairment and laryngeal pathology. This development has been separate from, and in conjunction with, the study of the speech system and speech disorders and has resulted in the need to address research needs and strategies in the speech and voice systems independently.

Definition of the Voice and Speech Systems and Their Disorders

Voice studies relate to the production of sound in the vocal tract, with emphasis on sound generation in the larynx. The respiratory system, which is an integral part of the voice system, provides the energy source and must be coordinated with glottic valving and supraglottic modulation of the glottal airflow. Vocal tract shaping above the larynx in the posterior pharynx, nasopharynx, and the oral cavity affects the quality of the voice, and is also part of the voice system.

Speech articulation involves kinematic adjustments of the posterior pharynx, tongue, velopharyngeal valving, and lip and jaw movements, which modify and enhance the sound source. Articulatory valving and obstructions of the airflow can produce significant downward modulations on glottic function. This is only one example of the close interaction between the voice and speech systems. Other significant interactions result from the effects of the tongue position on the height of the hyoid bone and larynx during speech. Therefore, although normal structure as well as diseases and disorders of the voice and speech systems can be studied independently, their interrelationships must also be studied.

High Priority Areas of Investigation

The following areas are identified as high priority and fruitful areas of investigation. They are classified on long-term and short-term bases. Short-term priorities are those in which existing instrumentation and basic knowledge can be applied immediately, with meaningful data emerging in a few years. Long-term priorities are those in which considerable background research is needed.

Long-Term Priority Areas

- Incorporation of molecular and cellular biology and anatomy to understand the microstructure of speech organs.
- Development of prostheses (laryngeal pacers, artificial larynges, etc.) for laryngeal insufficiencies.
- Development of surgical procedures in conjunction with predictive computer simulations.

Short-Term Priority Areas

- Muscle, reflex, and receptor physiology of laryngeal muscles (and other speech muscles), with specific emphasis on neural connections, neurotransmitters, and central control mechanisms. Anatomic and physiologic norms should be sought in these areas.
- Biomechanics of the larynx (skin, ligament, cartilage and muscle, and membrane tissue properties and their integrated mechanical actions).
- Critical periods of development of the phonatory system.
- Regulation of pitch, loudness, register, and vocal quality (normal and trained vocal production).
- Validation of animal models for phonatory processes.
- Deterioration of the laryngeal tissues and the pulmonary system with age; the deterioration of vocal control with age.
- Idiopathic voice disorders.
- Neurologic disorders of the larynx.
- Recovery from surgery, injury, and vocal abuse.
- Swallowing disorders related to laryngeal function.
- Development of techniques for kinematic, acoustic, aerodynamic, neurophysiologic, and visual observation,

including stress tests and diagnostic utterances.

Background

The larynx, or voice box, contains several cartilages that surround the top of the trachea. The cartilages have folds of tissue between them that are moved by muscles of the larynx. The folds are known as the vocal folds or vocal cords.

An important role of the larynx is life support. The larynx keeps the vocal folds open during breathing so that air can pass through the trachea to the lungs. During swallowing, the vocal folds must close tightly, and the larynx moves up behind the epiglottis, so that food will not enter the trachea to the lungs. During cough, which is a protective reflex, the vocal folds close and then open abruptly to expel air. In the process, food or liquid is prevented from entering the lungs. The larynx is an essential organ for maintaining the airway for breathing and for protecting it during swallowing. These actions prevent life-threatening aspiration pneumonia.

Voice, or phonation, is generated when airflow from the lungs is modulated by the vocal folds in the larynx. The vocal folds are brought together by the muscles and vibrate when air is pushed past them with sufficient pressure from the lungs. The vibration of these folds causes the airflow to become pulsed. This pulsed airflow is then modulated by the vocal tract structures (the pharynx, oral cavity, and nasal cavity) to generate sound waves, which are perceived as voice during speech. Without normal vibration of the vocal folds in the larynx, the sound of speech is absent and people can only "mouth" words. They cannot be heard or understood by others, either in face-to-face conversation or over the telephone. To produce a whisper, the vocal folds need to be partially closed, to cause a rushing of air that allows some speech sounds to be partially understood by persons very close by.

When vocal fold vibration is impaired, sound generation for speech is affected. An absence of one or both of the vocal folds results in a loss of voice, or aphonia. This happens when all or part of the larynx is removed, as in laryngeal cancer. Growths on one or both of the vocal folds can change the mechanical properties of the tissue, which affect its vibration. Growths such as laryngeal cancer or benign neoplasms such as polyps result in a hoarse voice due to irregular vocal fold vibration. Other tissue changes, such as nodules, edema, or contact ulcers, can also cause a hoarse voice. Papillomatosis, a spreading of wartlike growths covering the vocal folds, also interferes with voice production by limiting fold vibration and the intake of air through the larynx to the lungs.

The nerves controlling the motor and sensory functions of the larynx can be impaired through accidents, surgery, or viral infections. When the motor nerves on one side are affected, the muscles moving the vocal fold are paralyzed, and the fold cannot come close enough to the center of the larynx or be tensed enough to vibrate. In such patients, there is excessive air loss between the folds during vibration, which results in a breathy voice. When nerves on both sides of the larynx are paralyzed, the

muscles may not open the vocal folds properly for breathing. This happens in some neurological diseases (for example, multiple systems atrophy) or following surgery or trauma to the larynx.

Both bilateral and unilateral laryngeal nerve paralysis can also interfere with swallowing, since laryngeal movement is reduced and the trachea may not be closed. When sensation is affected, the coughing reflex does not function normally, and there is danger that food or liquid will enter the trachea, which can result in pneumonia.

Until recently, the only treatment for this condition was a tracheostomy, in which an opening is made in the trachea below the larynx to allow the air to enter the lungs. A tracheostomy interferes with speech since air is not passed through the larynx except when the opening is covered.

Laryngeal movement disorders have only recently been recognized. In spasmodic dysphonia, the muscles contained in the vocal folds move abnormally during speech, causing uncontrolled pitch and voice breaks and sometimes affecting swallowing and breathing. Vocal fold tremor, another disorder that is due to rhythmic contractions of the muscles of the larynx, causes the voice to shake with frequent pitch and voice breaks. Another disorder, laryngospasm, is an uncontrolled closing of the vocal folds, which interferes with breathing.

All of the voice disorders have devastating effects on the patient. They interfere with speech and oral communication, making the patient unable to function in most work situations. Use of the telephone is often impossible, and some patients are forced to rely on writing for communication. Many careers can be lost due to onset of a voice disorder. When respiration and swallowing are affected, life is threatened, and the only available treatment may result in a complete loss of voice.

RECENT ADVANCES

Photography of the Larynx

Until recently, examination of the larynx was by indirect laryngoscopy. In this procedure, the otolaryngologist places a mirror at an angle in the back of the patient's mouth to obtain a view of the vocal folds during breathing and during production of a short vowel. Many patients gag during this procedure, and usually only a brief view of the folds during phonation can be obtained. Further, since a mirror interferes with speech, vocal fold movement during speech cannot be seen.

During the last 10 years, videofiberoptic nasopharyngoscopy has been introduced. It is used by most specialists to make video recordings of the larynx and vocal folds during speech, swallowing, and breathing. A small tube is passed through the nose, and the end hangs over the larynx for viewing vocal fold movement. The patient can speak naturally during the examination, and the physician can view movements of the larynx and vocal folds for periods of up to 20 minutes.

Laryngeal stroboscopy employs strobed or paced bursts of light, which are synchronously timed with vocal fold vibration. The procedure provides a slow-motion view of vibrations of the folds during speech, which are far too rapid to be seen with the naked

eye. With the addition of laryngeal stroboscopy within the last five years, vocal fold vibration, ranging between 100 and 400 cycles per second, can be seen in slow motion and recorded on videotape in the laboratory or in the scientist's office. The extent of vibration of each fold and the degree to which the folds meet during vibration can be determined. Another tool, the rigid laryngoscope with magnification, is used to record on videotape the anatomical details of the vocal folds and laryngeal structures. An immediate recording and viewing of the larynx, vocal folds, and their movements during speech can be obtained. These techniques are essential for diagnosis. They provide a permanent record of a patient's problems, and the patient can view the record while the problems are explained.

The advances in video recordings of the larynx have led to improved understanding of laryngeal movement disorders, which can be seen only when vocal fold movement is observed during speech. Spasmodic movements, excessive vocal fold closing, opening, shortening, lengthening, tremor, and incoordination can be seen and recorded for diagnosing spasmodic dysphonia, vocal tremor, and laryngospasm. Previously, many of these disorders went undiagnosed, since the larynx could be examined only at rest.

Clinical use of laryngeal stroboscopy has led to improved understanding of voice disorders in the last few years. It has been learned, for instance, that for normal voice, the mucosal covering layer of each vocal fold must be free to move during vibration in a wavelike fashion. Any growth or scar that can interfere with the mucosal wave will result in an abnormally hoarse voice.

Acoustic Measurements

Listener impressions are very unreliable. Until recently, the otolaryngologist and speech-language pathologist were without methods of obtaining measurements of acoustic characteristics of voice disorders. Many noninvasive tools have been developed over the last 10 years for measuring voice and for diagnosing which aspects of phonation are impaired. Until very recently, these types of voice analyses were available only to voice scientists and required many months to produce.

Microprocessors can now automatically analyze voice signals and provide real-time displays of the frequency of vocal fold vibration and voice intensity. Other microprocessors provide real-time measures of the variability of the frequency and amplitude of voice signals (jitter and shimmer). Video sound spectrography provides a real-time spectral analysis of voice, and long-term spectral averaging provides an analysis of the energy contained in the voice signal at different frequency ranges during connected speech. Each of these measures provides objective means of measuring acoustic parameters that reflect voice disorders such as breathiness and hoarseness.

Measures of Vocal Fold Vibration

The next generation of measurement tools is now undergoing further development in voice laboratories. The tools provide physiologic information on vocal fold

vibration and airflow modulation. With the electroglottograph, scientists can now assess whether the vocal folds meet during vibration. With photoglottography, the area of glottal opening between the vocal folds can be measured. Airflow and air pressure devices are available for measuring the modulation of the air column by the vocal folds and also the amount of air pressure exerted from the lungs. These measures are being used by scientists to study how phonation is produced, and they will soon be used as diagnostic tools for studying laryngeal disorders. Scientists are also making recordings of the electrical activity of the laryngeal muscles in patients and in normal speakers to determine the role of each muscle in vocal fold opening, closing, and tensing for phonation. This technique is fast becoming a diagnostic tool for patients with vocal fold paralysis following injury to the laryngeal nerves.

The development and use of these tools by voice scientists are leading to changing concepts and new theoretical models of vocal fold vibration during the production of voice. Previous models of laryngeal vibration did not take into account flow dynamics, tissue strain, or the structure of the vocal folds. The importance of the viscosity of the mucosal layers covering the vocal folds and their different properties of movement from the underlying layers or body of the vocal fold is now recognized. The movement trajectories of the vocal folds are now known to be three-dimensional rather than only planar (lateral-medial plane). The stress-strain properties of these tissues are being measured to determine the biomechanics of vocal fold responses to airflow dynamics. These biological models have radically changed the understanding of how phonation is produced and of the properties that are essential for it. This knowledge will lead to the development of better laryngeal prostheses for postlaryngectomy patients.

Imaging the Larynx and Pharynx

Magnetic resonance imaging (MRI) is now being used to measure changes in the shape of the vocal tract. Such changes modulate the pulsed airflow produced at the larynx for different vowel sounds. Three-dimensional views of the contours of the pharynx, which have never previously been available, will provide improved understanding of how phonation becomes speech.

The importance of laryngeal movement during swallowing is being studied with video recordings of ultrasound to determine how movement trajectories are affected by disorders of the laryngeal musculature. Improved understanding of laryngeal dynamics required for the swallow will have an immediate impact on the diagnosis and treatment of a significant proportion of patients with swallowing disorders.

The Neurophysiology of Laryngeal Movement

Several voice laboratories are studying laryngeal muscle activations to determine the biomechanics of laryngeal muscle forces

to produce different movement trajectories during respiration, airway closure, swallowing, coughing, singing, and speech. Recently, properties of laryngeal muscles were found to differ from properties of other muscles. Laryngeal muscles are highly resistant to fatigue, are able to contract very rapidly, and are less susceptible to injury. Experimental studies have demonstrated reinnervation of the laryngeal muscles after nerve injury with and without surgical intervention.

The possibility of a neural prosthesis that can produce normal patterns of laryngeal movement in patients with bilateral laryngeal nerve paralysis is being tested in the laboratory. These experiments should lead to the development of microprocessors for controlling stimulation electrodes to open and close the vocal folds for breathing, swallowing, and speech.

Advances in the Diagnosis and Treatment of Voice Disorders

Vocal Fold Neoplasms

The development of acoustic measures of voice that can be used to screen persons for early signs of laryngeal disorders holds promise for improved early detection of persons at risk for laryngeal cancer. If such disorders are detected early, the prognosis for cure and maintaining voice function is very high. The development of improved visualization for the larynx and video recordings will also allow for improved accuracy in early diagnosis of laryngeal cancer.

Following removal of the larynx for treatment of laryngeal cancer, patients very shortly after surgery can now be fitted with prosthetic devices to produce voice. Previously, such patients had only two alternatives: to learn esophageal speech following many months of therapy, or to use an electrolarynx. The Blom-Singer and other prostheses provide voice for speech without training and without an external device. Shortly, refinements in design will make speech sound with these prostheses more natural.

Phonosurgery is a new specialty requiring skill and new instruments for delicate operations on the vocal folds. Surgical techniques are being designed to improve and restore voice by removing benign growths, correcting structural abnormalities, and repairing trauma. Dramatic improvements have taken place on how best to maintain normal vocal function. Previous common practices such as vocal fold stripping have been found injurious to the voice.

Contact ulcers of the vocal folds, previously thought to be due to excessive speaking, are now understood to result from esophageal regurgitation and can be treated effectively. Papillomatosis is now understood to be viral in etiology. With identification of the virus, new approaches can be developed for the treatment and prevention of this devastating disease.

Vocal Fold Paralysis

Unilateral paralysis (paralysis of one vocal fold) can now be corrected by a variety of new techniques that have a po-

tential for much better results and fewer complications than those that occurred previously with teflon. Surgical reinnervation is still somewhat experimental, but it holds excellent promise.

Bilateral paralysis of the vocal folds is life-threatening since the airway is no longer open for breathing. In some cases, surgical repositioning of one vocal fold can be used instead of tracheostomy, and it maintains voice.

Disorders of Vocal Fold Movement

Until 10 years ago, spasmodic dysphonia was thought to be a psychiatric disorder. Patients have frequent voice and pitch breaks, exert great effort in speaking, and often must resort to whispering to communicate. This is now recognized to be a neurological disorder affecting the muscles of the larynx, which can be diagnosed with videolaryngoscopy. For patients with excessive closure of the vocal folds and other structures, the only surgical treatment until recently was to cut one of the nerves to the larynx. The symptoms in the majority of cases, however, return within three to five years after surgery. Last year, a new experimental treatment using botulinum toxin was found effective. This toxin temporarily paralyzes one vocal fold so that the excessive closure is reduced and voice can be easily produced. Such patients are symptom-free for two to three months following an injection, and they continue to respond to repeated reinjections every three to four months.

RESEARCH STRATEGIES AND PRIORITIES

Normal Structure and Function

Laryngeal and Respiratory Physiology

Laryngeal and respiratory physiology represents the foundation of existing knowledge and serves as the basis for future investigations of voice production. Much present-day knowledge is organized on the basis of animal experimentation, which has limited applicability to human functions. Modern technology presents the opportunity to obtain direct insight into the specialized functions of the human larynx, with specific regard to receptor identification, reflex organization, central control, and muscle behavior in health and disease. This area of research represents a special opportunity for great and speedy advancement, and it is a high priority.

Studies are needed to evaluate the compartmentalization of the laryngeal muscles with regard to histochemistry and motor unit organization during different gestures, such as inhalation, exhalation, effort closure, phonation, and swallowing.

Information about the modification of sensory and muscle action feedback to the motoneuron pools during different laryn-

geal gestures is needed for higher level primates and for humans.

Recent noninvasive techniques offer new opportunities for research on humans, such as transcranial magnetic stimulation and sensory evoked potentials. The techniques can be used for mapping the cortical control of the motor and sensory aspects of the larynx.

Electrical stimulation of the recurrent nerve can elicit both short and long latency reflexes, and provide new insights into the on-line modifications of activity of the brain stem motoneuron pool during different phases of respiration and phonation.

Biomechanics of the Larynx

To understand how vocal pitch, loudness, and voice quality are controlled, it is important to study the mechanism by which the laryngeal and respiratory muscles move tissue and air. What kind of configurations are produced by groups of muscles working in concert? How do the elastic and viscous properties of various layers of vocal fold tissue affect vibration? Does transport of fluid into and out of laryngeal tissues restrict the range of vibration of the vocal folds? What are the properties of sound sources in the larynx and the vocal tract?

The biomechanics of these complex structures and material properties, which formerly posed many problems for quantitative analysis, can now be addressed by finite element and other computational means. In other fields of biomechanics, the mechanical properties of skin, cartilage, fat, ligament, tendon, and muscle have now been studied extensively. The results give voice researchers a rich data base for comparisons. This combination of new tools of analysis and new data bases makes this a high priority area of investigation. It is likely to lead to significant progress in a few years and lay the groundwork for simulation of reconstructive and augmentative clinical procedures.

Cellular and Molecular Biology and Anatomy

Normal laryngeal anatomy has been studied in many species including human postmortem specimens on both light microscopic and ultrastructural levels. These studies, using baseline information, provide an opportunity to define the distribution of extracellular matrix molecules, oncogenes, growth factors (for example, epithelial growth factor), and growth-factor receptors as they relate to normal structure of the larynx. A vast body of current literature demonstrates that the distribution of these factors in the intact normal larynx and their interactions are essential for the maintenance of normal stable conditions. Shifts in the relationships of these components have been implicated both in disease processes and in wound healing. New information can be developed with existing cellular and molecular biology techniques of immunocytochemistry and in situ hybridization. Acquisition of this information is an important long-term goal.

Regulation of Pitch, Loudness, Register, and Vocal Quality

Over the past decade, functional models have enhanced the knowledge of the

dynamic behavior of vocal folds. Electromyographic studies have increased the understanding of the role of the extrinsic laryngeal and the cricothyroid muscles in the regulation of pitch. The results of the studies have suffered, however, from the small populations sampled and from individual variations. Motor equivalence for phonatory tasks, for example, is still unclear. Systematic work on the regulation of pitch, loudness, register, and quality should be continued. Concentrated work is needed on the role and interactions of specific muscles and of their aerodynamic properties in relationship to changes in the length, shape, configuration, and forces of the vibrating structures. To address these concerns, studies are needed using models, live speakers, autopsy material, and premorbid recordings.

Perturbation Analysis

Acoustic analysis of the human voice to determine neurologic, respiratory, and laryngeal disease is entirely noninvasive and economic, and it is constantly enriched by new signal processing techniques. A major goal in acoustic analysis is to identify the quasiperiodic characteristics of the source of sound. Because the vocal fold-vocal tract system constitutes a complex biological oscillator that can reflect many disorders of the body, much information is expected to be gained from a formalized study of its perturbations. Recent improvements in the theory of chaos (a new field of study that attempts to find order in seemingly random behaviors) might be combined with more traditional nonlinear system analysis to describe irregularity in vocal fold vibration. The results may lead to a better detection of specific disturbances, such as asymmetry in vocal fold structure, excessive mucus on the vocal fold surfaces, randomness in neurologic input, and aerodynamic instabilities.

Source-Tract Interactions

The interactions between phonation, respiration, and articulation are significant. Research should stress the importance of the pulmonary system as an energy source for the voice. The average person uses only 1 to 10 percent of the total available respiratory air for speech. Additional demands such as increased loudness may add additional loads to the system. Here, the neural reflex systems must support requirements for additional air. Although such demands have been studied, they must be much more clearly defined for different types of vocalization. In addition, the role of changes of supraglottic pressure (acoustic and aerodynamic) in vocal function and training could be better understood if the neural sensors that help control this process were more clearly defined.

Acoustics and biomechanical interactions should also be studied further, particularly the effects of vertical movements of the larynx and source-filter effects related to adjustments in the lower pharynx.

The dynamic behavior of the larynx and its role as an articulator in speech need further attention. Studies focused on the regulation of timing of laryngeal behavior in coordination with respiratory and articulatory activity can shed considerable light on communication disorders.

Vocal Fatigue

Increased daily usage of vocal communication and increased environmental noises have resulted in increased demands on the vocal system. Complaints of vocal fatigue by teachers, public speakers, coaches, salespersons, and many other professional and nonprofessional users of voice are common. When fatigue occurs, it is important to know the degree to which central and peripheral mechanisms are involved. At the periphery, the contractile mechanism of a muscle may fatigue, or nonmuscular tissues (ligaments, joints, tendons) may lose their ability to support mechanical stress. Recovery periods for these processes may differ, and a clinician's advice to a patient may depend on the type of short-term or long-term recovery needed. Central fatigue, or the inability of the brain to continuously activate the motor system, may also play a role, but perhaps to a lesser degree. An opportunity for research exists, especially in light of similar problems being encountered by athletes and performing artists who place heavy demands on parts of their bodies. Sports medicine is a growing field, and the proper maintenance of the body under unusual mechanical loads, whether they are on a limb or the larynx, is a health concern. Research on vocal fatigue may also help retard or prevent the "geriatric voice" and increase vocal skills and longevity in professional speakers.

Role of the Larynx in Swallowing

Little attention has been given to the motor control of the larynx in the swallowing process. The timing of the laryngeal elevation, protective closure, and esophageal-opening actions of the larynx need to be determined. Experimental studies to define the actions required of intrinsic and extrinsic laryngeal muscles should provide a significant new understanding of the role of the larynx in swallowing.

Neural Regeneration

Paralysis of the vocal mechanism simultaneously disrupts its phonatory, protective, and respiratory functions. Attempts to artificially reposition paralyzed folds unfortunately trade one function for another. Ideal therapy consists of reinnervating paralyzed muscles to restore appropriate vocal fold dynamics. Knowledge of neural regeneration is a key to an ability to restore complex function after denervation.

Development

Validation of Animal Models

Most of the information currently available on the neurological control of the larynx has come from animal studies. Recent clinical studies of laryngeal reflexes suggest that the neuroanatomical and neurophysiological control of laryngeal functions in humans may differ from that of many other species. Attention needs to be given to the similarities and possible differences in human laryngeal neurophysiology in comparison with that of other species proposed as models for human voice production. The validity of the study of

laryngeal neurophysiology in higher level primates should be explored. This research is fundamental to an understanding of developmental structure and function and has a high priority.

Embryology

Current knowledge consists of a series of well-defined descriptive reports of different stages of laryngeal development in several species, including humans. Precursors of the larynx consist of two cell populations that must interact numerous times during the developmental process to form the adult organ. As yet, no reports address the mechanisms (epithelial-mesenchymal tissue interactions) that are the driving force behind normal development of the larynx, and the way that these interactions affect cell differentiation and final organotypic form. The understanding of the embryology of this unique organ is, therefore, still quite limited, as well as the understanding of repair processes in the injured larynx and after reconstructive surgery. Studies that address developmental mechanisms should be favored over those that propose purely descriptive work.

Comparative Anatomy

Since Victor Negus' comparative anatomy of the larynx and supralaryngeal pharynx approximately 40 years ago, there have been few contributions in this area. For future research, detailed comparative descriptions are needed that could be used to facilitate the selection of models appropriate to human function and reduce the number of animals needed to address issues concerning laryngeal function.

Critical Periods

Critical periods for optimum development of vocal control as well as periods when a speaker may be at risk for developing dysphonias have been recognized, but they have not been adequately studied. The critical periods identified as particularly important for prevention and treatment of voice disorders include: infancy, puberty, auditory impairment (if applicable), and periods associated with hormonal change. More specifically, the periods of importance are:

- Infancy. A time when vocalization is often at loud levels for long durations, or when vocalization is playful.
- Puberty. A time when there is a rapid growth in the larynx resulting in significant vocal fold lengthening with perhaps insufficient concomitant time to allow for proper mechanism adjustment for many adolescents. That this adjustment cannot be done readily results in abnormal attempts to maintain the higher childlike voice at the expense of vocal strain and poor vocal quality. These vocal characteristics may result in unwanted attention and have a significant effect on the psychosocial development of a speaker. A better understanding of the mechanism during puberty is needed, along with bases for therapy.

- Hormonal changes. Times when masculinizing tumors or male hormone therapy, pregnancy, or menopause result in edematous changes in the laryngeal structure. As yet, there are no studies to determine the time periods at which the problem is reversible after onset. Research is needed to enhance the understanding of the androgenic hormone and how its effects can be prevented and treated.

Vocal Control

Some changes in the laryngeal system co-occurring with development have been well documented. Research studies are needed to characterize voice production as it relates to the developing structure. A major concern is the regulation of pitch, loudness, register, and vocal quality with changes in morphological structure. Knowledge of normal structure and function relationships as a function of age is requisite for understanding voice production, for developing standards of tests for early detection of disorders, and for determining the efficacy of various forms of treatment. This information is also valuable in developing theoretical models of the etiology of voice disorders. Vocal control is a high priority for further study.

Effects of Deafness on Developmental Vocal Control

Individuals with either congenital deafness or acquired hearing impairments have disorders of frequency and loudness control as well as distinctive abnormalities in voice quality. Only limited attention has been given to the critical periods for the habilitation of vocal control in the congenitally deaf. Visualization of the acoustic signal has not been a successful method of voice training. Attention needs to be given to the use of physiological feedback and motor learning in training vocal control in the congenitally deaf and in adults who are profoundly deaf.

Aging

Preliminary observations indicate that some of the vocal alterations that have been ascribed to "aging changes" are perhaps reversible and are not due to irreversible physiology of anatomic alterations. More research is needed to establish the true nature of geriatric voice changes and to develop techniques to forestall or prevent them when such changes may interfere with a person's life or livelihood. In addition, the irreversible changes need to be understood in depth.

Cell Death

The possibility of selective neuronal cell death in the brain stem as a consequence of normal aging should be addressed. The answer may provide important insights into understanding reduced airway protection and voice control in older persons. In some neurological diseases, selective reduction in motoneuron pools that modulate breathing and speech have recently been found. Research is needed to determine if selective

cell death is also part of the aging process and adversely affects respiration and voice.

Anatomical, Cellular, and Molecular Biology

The anatomy of the aging larynx has been described in human specimens obtained primarily from cadavers. There has been no detailed study of the aging process in other species where a more uniform genetic background is possible. It will be very useful to collect anatomical, cellular, and molecular information from aged larynxes of a genetically defined species. Specifically, information on the localization of extracellular matrix components, growth factors, oncogenes, and growth-factor receptors is needed. This information should be compared with the data collected from normal adult larynges and related to reduced ability of the larynx to recover from injury with aging.

Muscle Atrophy

It is well known that muscle tissue degenerates with age. Muscle fiber degeneration usually results from loss of innervation. Sometimes reinnervation from larger and healthier motoneurons causes the muscle tissue to bunch into larger fascicles. The combination of fiber bunching and total loss of fibers causes overall weakness and reduced stability of muscle contractions. This state is likely to be reflected in reduced ranges of pitch and loudness, in inability to regulate the breath stream, and in increased unsteadiness of the voice. Further research is needed to determine the acoustic consequences of muscle atrophy in the laryngeal and respiratory systems.

Deterioration of Joints, Ligaments, Membranes, and Other Tissues of the Laryngeal, Pulmonary, and Secretory Systems

Vocal deterioration involves restrictions in the movement of the glottic structures as well as reductions in lubrication. Specifically, the cricoarytenoid joint responsible for the glottic abduction and adduction capabilities tends to lose the freedom of glide and turn because the capsule of the joint itself deteriorates. Further, arthritic changes can even result in a joint that is fused in a fixed position. Little is known about the biology of this process. To correct these changes by means other than surgery, more information is needed about the cell biology of joint deterioration in the aging larynx. This knowledge might lead to medical, exercise, and perhaps nutritional therapy to help arrest or reverse the process.

Stiffness and lack of stretch in the ligament structure contribute to a less pliable organ. This state affects the membranous cover, which results in less motion of the mucosal wave. When the system is less capable of driving the glottis and vocal stress is increased, support requirements of the pulmonary system are increased.

Further increased stiffness is associated with less mucosal secretion and a tendency

toward dryness, which makes clear phonation more difficult in aging. Research is needed to determine how to improve the secretory process leading to increased pulmonary efficacy.

Exceptional (Trained) Vocal Behavior

In normal conversational speech, the larynx is a very inefficient sound source. Typically less than 1 percent of the aerodynamic power of the lungs is converted to acoustic power in speech. As a result, the system can become "lazy," or it might optimize itself for criteria other than efficient phonation. Undesirable vocal habits may be formed in various developmental stages.

When people with poor vocal habits are thrust into situations where prolonged periods of vocalization are required, especially with high vocal intensity under high psychological or physical stress, this loss of efficiency takes its toll. Vocally trained people (singers, actors, public speakers) have learned methods to overcome some of these problems.

The gifted or exceptional vocalist, either in speaking or singing, offers a very important model for vocal evaluation of all voice users. The model establishes the limits of capability. In pitch control especially, it allows one to see what can be done with respect to range. This information can be used in average voice evaluation as both a criterion and a role model. The same approach is applicable with respect to loudness and maximum phonation time. Questions of interest are:

- How far can the human voice be pushed without strain? What are its limits? Answers to these questions could lead to better care of people who have deficient usage.
- What is the basis for limited efficiency? Is it genetic? How is optimal efficiency achieved?
- How does the optimal user gain a greater range of pitch or loudness? The answer to this question would lead to better care of a person with a narrow vocal range and low power. It is important to know the period of training required for near optimal usage.

Results of the study of pitch, loudness, and vocal quality control of this population will help speech-language pathologists and otolaryngologists remediate vocal disorders, particularly in the direction of modifying behavior. This is considered a high priority area of study for the near future.

Diseases and Disorders

Structural

Congenital and Acquired Laryngeal Pathology

Congenital and acquired lesions of the larynx result in blockage of breathing in thousands of infants and children each year. These disorders may occur as cysts, webs, or scar tissue near the vocal folds, or they may result from failure of the larynx to develop a strong supporting skeleton. The latter situation can result from interruption

of epithelial-mesenchymal tissue interactions during organogenesis of the larynx. Presently no animal models exist to aid investigators in their study of these developmental problems. Identification of genetic and environmentally induced laryngeal anomalies is an important consideration and is essential for improving the understanding of the mechanisms that are active in the cause of these pathologies.

Inflammatory and Infectious Diseases of the Upper Airway and Larynx

A high priority should be given to research to improve the understanding of inflammatory diseases of the upper airway and larynx. Rhinitis, sinusitis, and laryngitis affect a high percentage of the population, and these conditions range in severity from local discomfort and blockage of breathing to chronic infection and tissue changes that cannot be reversed without surgical therapy. Acute and chronic infection of the upper airway may result in systemic illness, fatal airway obstruction, and infections of the central nervous system. Results of research on prevention, early detection, and rapid intervention should help to limit the effects of these processes. Newer endoscopic sinus surgical techniques appear to represent a significant technical advance, but studies are needed to accomplish a reduction in the associated complication rates and an enhancement of long-term therapeutic benefits. Research designed to produce new medical treatments for infectious and allergic disorders of the upper airway and larynx should also be emphasized.

The incidence of acquired laryngeal papillomas in young adults appears to be increasing. Their management is especially perplexing in professional voice users. A clearer understanding of whether they are benign or potentially premalignant lesions would help in determining a therapeutic course. It has been well established through gynecologic research that specific viral types of papillomas are associated with the development of uterine cervical carcinoma. Long-term, interinstitutional research should permit viral typing of laryngeal papillomas and assessment of their recurrence rate, effects of treatment, effects of cofactors such as smoking and voice abuse, and rates of association with carcinoma of the larynx.

Tumors of the Upper Aerodigestive Tract

Tumors of the oral cavity, pharynx, larynx, and neck may result in loss or serious impairment of the ability to communicate with others. Benign tumors such as recurrent laryngeal papillomata may require multiple surgical (usually laser) excisions from the vocal fold region. Postoperative scarring may lead to extreme hoarseness, while failure to control these tumors in some cases results in death. Clinical trials with interferon have shown that laryngeal papilloma responds to immunotherapy, but the response is only temporary. Studies are needed of prevention, early detection methods, and improved surgical and immunologic treatment of these patients.

Malignant tumors involving the larynx and other head and neck structures affect more than 50,000 Americans each year. Cancer in these structures is usually related

to the use of tobacco and alcohol, and research into more effective cessation strategies is needed. Early diagnosis often permits the use of limited surgical excision, such as partial laryngectomy or partial glossectomy. While these operations conserve most of the physiological functions of speech, respiration, and swallowing, more precise data are needed on the long-term deficits that are produced and on strategies to rehabilitate patients. Standardized, prospective clinical investigation and the collaboration of surgeons with speech scientists must be expanded in this area. The technology and expertise exist to accomplish major advances in this field within a five-year period.

Neural

Vocal Fold Paralysis

Vocal fold paralysis most often occurs unintentionally during surgery of the skull base, neck, or mediastinum. Tumors or infections along the path of the vagus nerve can also interrupt laryngeal function, producing alternations of phonation, airway protection, and respiration. In bilateral paralysis, the consequences are often devastating. To date, observations about the effects of laryngeal denervation remain controversial. Two opportunities for urgent research must be addressed. Accumulation of a sufficient data base will improve the ability to diagnose accurately and treat effectively these life-threatening conditions. In addition, multi-institutional clinical trials are required to assess existing and new surgical procedures including vocal fold augmentation, thyroplasty, arytenoidectomy, cordectomy, selective reinnervation, and electrical pacing. Major advances are expected to be achieved over the next three years. This category of research is a high priority.

Spasmodic (Spastic) Dysphonia

Significant new advances have been made in understanding the idiopathic disorder of spasmodic dysphonia. Neurophysiological study of the intrinsic laryngeal muscles has demonstrated that this is a focal dystonia affecting the laryngeal musculature during respiration, speech, and swallowing. Brain imaging techniques have indicated that in some cases the etiology of the disorder can be due to discrete brain lesions in the motor tracts. Further major advances can be expected in the next three years in understanding the pathophysiology of this disorder. Epidemiological and case control studies are needed to identify possible etiological factors leading to the onset of this disorder in midlife.

Vocal fold tremor affecting activity in the laryngeal muscles during exhalation and voice production has recently been identified as a focal motor control disorder frequently isolated to the larynx and appearing in midlife. Significant advances in determining the pathophysiology of this disorder can be expected in the next three years.

A significant advance in the treatment of both spasmodic dysphonia and vocal fold tremor has recently been reported. Minute injections of botulinum toxin into the intrinsic laryngeal muscles significantly reduce the symptoms in most cases for three to four months. Long-term evaluation of this treatment is needed to determine whether it

continues to be effective with repeated injections over several years.

Laryngeal Disorders Secondary to Neurological Disorders

Control of the laryngeal functions both for breathing and voice are impaired in many of the neurological disorders such as Parkinson's disease, amyotrophic lateral sclerosis (Lou Gehrig's disease), multiple systems disease (Shy-Drager syndrome), and Huntington's disease. Studies of the neural control of the larynx are needed to determine why laryngeal functions are particularly vulnerable to neurological disorders causing selective neuronal loss either in the brain stem or in the basal ganglia. A better understanding of the pathophysiology of these laryngeal motor control disorders would lead to more effective management (medical, surgical, behavioral, or prosthetic) to support respiration and speech. The techniques and investigations currently ongoing or forthcoming in this area suggest that significant advances will be made over the next three years. These studies should provide badly needed new knowledge about neuropharmacologic and behavioral management of disorders of laryngeal motor control.

Some studies are needed to clarify the relationships between neuropathology and laryngeal disorders. Information about characteristics accompanying different neurologic disorders and disease progression are lacking.

Idiopathic Dysphonia

Idiopathic vocal disorders are those disorders in which the laryngoscopic anatomical appearance is normal and the vibratory pattern is abnormal for no discernible reason. These problems occur with considerable frequency, and they represent a source of great frustration for patients and therapists alike. Idiopathic dysphonias are often resistant to treatment because the underlying pathophysiology is not understood. Frequently, inappropriate clinical decisions are made despite recent advances in the diagnoses of many other voice disorders. Knowledge about the mechanisms underlying the formation of such dysphonias is vitally needed. Causative factors that may adversely influence voice production, such as environmental noise and air pollution or systemic disease, need to be examined.

Environmental Factors

There is a national need for research to expand the knowledge of the effects of environmental agents on the upper respiratory tract (nose and sinuses) and the larynx. Humidity, excessive dryness, smoke, fumes, dust, and pollen are known to affect adversely the normal functioning of these organs. These environmental factors are implicated in the long-term development of nasal and sinus infections, sore throat, hoarseness, and even cancer. Long-term exposure of a person to workplace toxins can cause both short- and long-term loss of time from the job. Particular areas of opportunity include epidemiologic surveys, clarification of the precise etiology and pathophysiology, and strategies for prevention, diagnosis, and management.

Recovery From Surgery and Injury

Wound healing in the injured larynx and postsurgical therapy can have a profound effect on the functional recovery of this generator of sound. Exciting new research advances demonstrate the role that some extracellular matrix proteins (for instance, fibronectin and its integrin receptor molecule) play in the improvement of the quality of wound healing. Recent studies demonstrate how wound healing can be accelerated and scarring reduced by the introduction of specific extracellular matrices. Myofibroflasts have also been implicated in scarring and contracture of healing wounds.

There is at present little knowledge concerning the role of extracellular matrix, growth factors, and oncogenes in wound healing in the larynx. Studies of these factors in wound healing in the injured larynx of animal models are essential to the design of future experiments, where specific treatments may be assessed that could reduce the negative effect that wound healing has on functional recovery.

The various strategies used in clinical practice to control or influence wound healing must be defined more clearly. Dilatation techniques, the use of lathyrogenic drugs, the timing of procedures, and the use of stents to support the airway during healing lack standardization. Basic science and clinical information are needed about the pathophysiology, prevention, assessment, and treatment of problems associated with laryngeal surgery and injury.

Endocrine and Pharmacological Voice Changes

There has been considerable scientific interest in the effect of hormonal and pharmacologic substances on the voice. These chemicals have a potent impact upon the distribution of electrolytes and water within the cellular compartments of the vocal folds, causing them to become edematous, or swollen, and changing the characteristics of their vibrations. These changes occur during hypothyroidism, premenstrual (edema) syndrome, androgenic hormone-producing ovarian tumors, pregnancy, and male puberty. They may also follow the administration of birth control pills and also postmenopausal hormone therapy in women. Studies are needed to clarify the best strategies for evaluating and treating these patients and to find new strategies for prevention. In addition, epidemiologic studies, standardization of clinical assessment, and a coordinated effort to establish and clearly define a data base in this area should be supported.

Respiratory Tract Disorders

The lower respiratory tract is an important, integral element in the production of the sound wave for speech. A variety of diseases and disorders affect the tracheobronchial tract, including neural and neuromuscular disorders such as cerebral palsy, head injury, and Parkinson's disease. Respiratory dyskinesia is a problem that has recently been differentiated as a separate

entity, sometimes associated with asthma. It presents an interesting opportunity for investigation because some patients can be successfully managed using anticholinergic drugs.

Disorders Secondary to Other Disorders

Other conditions of importance include infections (viral, bacterial, and fungal), inflammatory diseases (allergic, collagen), emphysema, and disorders secondary to trauma or inhaled toxic substances. More information is needed concerning the effects of reduced respiratory capacity on speech and on the neuromuscular mechanisms and reflexes regulating respiratory volume and flow during speech. New developments in endoscopy have expanded the capacity to observe and record information, which should enlarge the understanding of the pathophysiology, diagnosis, treatment, and rehabilitation of these patients.

Dysphagia

Abnormal swallowing can produce devastating consequences, ranging from interference with nutritional support to causing life-threatening aspiration pneumonia. The frequent co-occurrence of selective voice and swallowing impairments in disorders such as spasmodic dysphonia, vocal fold paralysis, and neurological diseases have implicated laryngeal motor control disorders in swallowing. Human and animal studies of selective intrinsic laryngeal muscle denervation are needed to gain improved understanding of the potential role of laryngeal impairment in many cases of dysphagia. Further, studies of the role of the brain stem nuclei in both laryngeal control and the swallowing process have significant potential for improving the understanding of dysphagia and identifying potential new approaches for treatment. Studies of the efficacy of treatment are especially encouraged.

Techniques, Procedures, and Devices for Analysis and Treatment

Diagnostic Aids

During the last decade, significant progress has been made in the development of new research tools for observation and quantitative measures of voice production. Systematic investigation of voice production using kinematic, acoustic, aerodynamic, and imaging diagnostic aids needs to be continued to determine which technique (or combination of techniques) is most useful in detecting disorders, documenting changes resulting from treatment, identifying persons at risk for developing voice disorders, setting standards for routine assessment, and improving the understanding of the process of voice production.

It is essential for research and clinical purposes that valid, reliable methods for objective voice measurement be established, including (at a minimum) measures of vibratory function, acoustic measures, phonatory function analysis, airflow, improved

electroglottography, and repeatable psychoacoustic measures. At present, there is little standardization of equipment. Definition of testing procedures varies from center to center, and no standards have been established for voice assessment. Objective voice measures should be as routine as audiometry is for hearing measures. Substantial research is needed to identify appropriate tests and develop a convenient, inexpensive armamentarium of equipment for performing the tests so that they may be used by practicing otolaryngologists and speech-language pathologists, and, to some degree, in conservatories and acting schools.

In addition to developing new equipment and standards for voice measures, previous studies that establish normative values have not been confirmed. Some of them were performed on small numbers of patients, and usually nonprofessional speakers were studied. More normative values are needed for the general population, professional speakers, and professional singers. Age and sex should be considered in all groups.

Electromyography

Although limited in its applicability because of its invasive nature, electromyography (EMG) is now considered an essential diagnostic and research tool. EMG has enhanced the understanding of vocal control and has helped to diagnose laryngeal paralyses differentially. It is essential to obtain further EMG data during different vocal control tasks, on various neurogenic based dysphonias, and in conjunction with other techniques. This information will play a central role in enhancing the understanding of pitch and loudness control, vocal fatigue, and knowledge of the etiologic bases of selected pathologies, and in determining if less invasive procedures can be used to provide diagnostic determination of the status of the recurrent laryngeal nerve.

Acoustic Analysis

Recently, manufacturers have begun to offer a wide variety of relatively low-cost acoustic analysis devices designed to provide quantitative information on voice production. In most cases, the diagnostic relevancy is still in question. Studies are needed of large samples of normal speakers at different age levels to establish a normative data base. Parallel studies of major categories of voice disorders are also needed to determine if sufficiently distinctive profiles make it possible to use this tool for detection and characterization of specific disorders.

Aerodynamic Measures

Measures of air pressure and airflow have been used to document changes resulting from treatment. The measures must be interpreted cautiously because of the effects of different levels of effort, potential influences of differences in pitch and loudness, and unknown sources of variability. Studies are needed to determine how these measures are affected by different pathologies, vocal fatigue, test tasks, changes in vocal control strategies, glottal configurations, and other aspects of vocal function.

Imaging

In the last few years, there have been several technological advances in the area of diagnostic imaging. High-speed computerized tomography (CT) scanners may make it possible to obtain cross-sectional shapes of the vocal tract. This capability should improve the understanding of supraglottal influences on voice production. It is now possible to use noninvasive ultrasonic technology to make longitudinal scans of the vocal folds in vibration. MRI provides the additional capability of viewing both source and vocal tract shape in the sagittal plane. This provides a major advantage for understanding vertical relationships during sustained vocalizations. It remains to be seen if this information will be useful for diagnostic purposes. The use of standard and stroboscopic light sources in conjunction with fiberscopes have demonstrated potential research applicability in studies of laryngeal movements and pathology. Such imaging techniques should be used in conjunction with other measures to help relate the structure to dynamic function of voice production.

Observations made from a combination of experimental techniques should be related to each other through appropriate models for understanding voice production and for application to clinical populations. Equally important to the specific technology is the development of test protocols that allow investigators and clinicians to make inferences about the flexibility and range of movements, breakdown in performance, and other factors pertinent to the development of dysphonias.

Prostheses

Paralysis by stroke, tumor, or infection greatly alters the ability of the larynx to perform its vital functions of airway protection, respiration, and phonation. The valvular activity of the larynx in each of these functions requires instantaneous alteration of vocal fold shape and position. For example, in breathing, the folds must abduct or lateralize to admit air. However, in swallowing, they must assume a closed position to block food from entering the lungs. During vocalization, muscles actively position the vocal folds near each other while altering internal tension and mass in order to effect precise pitch changes. No single reconstructive surgery can restore all functions. It often trades more of one function for less of another. Advances have been made in the development of electronic artificial larynges (external, internal, and implantable) and in tracheoesophageal shunts (with and without alloplastic voice prostheses). This area deserves support for further studies. New technologies in signal processing now allow the possibility of precisely activating individual laryngeal muscles at the precise moment that they are needed for subserving the diverse requirements in speaking, swallowing, and breathing. Several laboratories now have the capability to electrically pace the paralyzed vocal fold to reestablish its breathing function. Unfortunately, these investigations have been sporadically reported, and many experiments were largely uncontrolled. For scientists to extend electrical pacing to a greater complement of individual muscles in the larynx, they must

systematically define the best electrode configurations, to give consistent and optimum stimulation strategies. Basic knowledge related to the physiology of muscle denervation is generally limited, and is unstudied in the larynx. Little is known, for example, about the effects of electrical stimulation on the preservation of muscle integrity known to degenerate when separated from its nerve. In many respects, laryngeal investigation could serve as a model for other medical scientists involved in electrical pacing.

More specifically, this research offers the immediate possibility of a breakthrough in treatment of the paralyzed larynx. On the basis of medical need and availability of new technologies, this category of investigation has a high priority. Major results could be expected in three years.

Surgical Procedures

Recent developments in surgery encouraged by new technologies such as the operating microscope and laser have given the surgeon an unprecedented ability to alter structure and, as a result, to improve vocal function. Phonosurgery, designed to alter vocal output, focuses on endolaryngeal as well as framework modifications to change fundamental frequency and/or intensity and to reduce perturbation by restoring symmetry. New laryngeal surgical techniques seek to modify vibrating mass, alter vocal fold tension, and reposition paralyzed folds.

To expand use of phonosurgical methods, two parallel developments are required:

- Standardized measurements of vocal output before and after surgery.
- Simulation modeling to predict vocal outcome before surgical manipulation.

These developments will elevate phonosurgery from art to science and encourage its wider application in the treatment of many vocal disorders. The efficacy of newer and apparently better phonosurgical procedures such as thyroplasty should be established objectively.

Surgical excision of the thyroid gland (thyroidectomy) and some other neck operations are associated with a risk of inadvertent injury to the superior or the recurrent laryngeal nerves. This type of injury produces various forms of laryngeal paralysis and subsequent changes in voice, respiration, and swallowing. Early efforts have been made to develop a system for intraoperative monitoring that will alert the surgeon if these nerves are stimulated during surgery. Further efforts in this area should be supported.

It is recognized that simulation of movement of complex biomechanical structures requires extensive computation. Resources should be provided to make it possible for competent scientists and engineers to access large fast computers to make the simulations highly interactive and close to real time.

Research in this category is expected to result in considerable advances and has a high priority.

Medical and Behavioral Treatment

The etiological bases of voice disorders and their subsequent treatment are varied. Treatment, which includes both medical and

behavioral management, is reportedly effective for some voice problems and ineffective for others. Data on the efficacy of treatment are in their infancy. There is little systematic knowledge of selection criteria for specific treatments, frequency or duration of treatment, factors contraindicating treatment, and longitudinal results. In addition, the results of early versus later intervention are not sufficiently clear. Substantial improvements can be achieved during the next three years through careful experimental identification and evaluation of existing and new techniques. Comparative effectiveness of techniques, as opposed to no treatment or a combination of techniques, needs to be studied.

There is a group of dysphonias that result from incomplete closure of the glottis. Medical treatment for these dysphonias includes surgical procedures that reduce or eliminate the glottal gap. Of particular interest are medialization and augmentation procedures. There is a need for well-controlled studies that document the effects of these procedures on voice production to determine which cases can be helped and which may not be improved. Results of these studies will provide a basis for further refinement of medical and behavioral management.

Significant new treatment advances have developed in the last two years using botulinum toxin injections into the laryngeal muscles for reductions in abnormal patterns of laryngeal muscle activation. Such treatments are effective in reducing spasmodic bursts and tremor in both injected and non-injected muscles in the larynx and upper vocal tract. It is expected that further refinements in the use of this treatment will be made in the next three years.

Many patients with spasmodic dysphonia have had symptoms return following recurrent laryngeal nerve resection. Several laboratories have demonstrated that the condition is associated with nerve reinnervation resulting in restrictions in vocal fold movement due not to paralysis but possibly to abnormal muscle activation by synkinesis of autoagonistic muscle groups. Botulinum toxin injections on the side of nerve resection are effective in reducing symptoms of spasmodic dysphonia in these patients as well. Further advances can be expected in the combined use of medical and surgical treatments in patients with idiopathic disorders of this type.

Pharmacologic management of patients with voice disorders has not been carefully explored, but long-term opportunities can be expected in the manipulation of endocrine and neurotransmitter levels. Potentially such interventions may control voice changes associated with pregnancy and menopause. In motor control disorders, anticholinergic, dopaminergic, and other agents have already been shown to have significant effects on abnormal muscle activation patterns. Systematic investigations of the use of such agents with objective measures of vocal function hold long-term promise and should be initiated.

The efficacy of various therapeutic techniques for professional and nonprofessional voice users should be established. It is suspected that some techniques (such as forceful push-pull techniques) that may have a place in the armamentarium of speech-language pathologists may be

particularly deleterious when used for common problems of the professional singer. The technique and efficacy of various therapeutic methods should be assessed on a scientific basis.

It is becoming clearer that selected techniques used by singing teachers may be extremely valuable in voice rehabilitation of speakers. Although these techniques are outside the realm of traditional speech-language pathology, close cooperation among numerous voice therapy teams around the country and increasing cooperation between the American Speech-Language-Hearing Association (ASHA) and the National Association of Teachers of Singing (NATS) suggest that interdisciplinary investigation may result in improved therapeutic methods. These methods should be formally assessed. In addition, research should be directed specifically toward rehabilitation techniques for voices following acute injury and surgery.

Training Needs in Voice and Voice Disorders

Assessment and treatment of individuals with voice disorders require an interdisciplinary approach. Currently, there are only a few centers where a patient could be expected to be seen by a knowledgeable voice team. This sparcity causes inappropriate diagnosis and treatment, and it delays incorporating new knowledge into clinical practice. Changes in training are needed for speech-language pathologists, otolaryngologists, voice scientists, and voice teachers. In addition to traditional education, clinical training should include an interdisciplinary approach that incorporates basic voice science, pathophysiology, and surgical and behavioral management. It should also include information and skills acquired by voice teachers who train professional voices. Accomplishing this goal will require additional education and training of educators in university and medical centers that provide training in these disciplines. (For further discussion of this problem, see chapter 7, "Research Training and Research Personnel Resources.")

The significant developments in voice sciences and clinical advances in the diagnosis and treatment of voice and laryngeal pathologies in the last decade have resulted in a significant need for continuing education of practitioners in otolaryngology and speech-language pathology. Many current practitioners received their professional training before the explosion of new knowledge in the voice sciences. For otolaryngologists, continuing education programs in the new diagnostic techniques and phonosurgery are needed to assure the transfer of the new knowledge into the clinical delivery system. Similarly, only a small number of speech-language pathologists have specialized in the diagnosis of voice disorders and voice therapy. Specialized continuing education in the voice sciences and workshops led by master clinicians are needed to meet the clinical needs for the transfer of new knowledge from the research laboratory to the clinic. In addition, the substantial knowledge base acquired by voice teachers in music and drama schools in the area of voice training should be tapped and integrated into methods for vocal rehabilitation.

Research and Training Centers

Many different disciplines are required for comprehensive study of voice problems and management of voice disorders. Recently, otolaryngologists, speech-language pathologists, neurologists, engineers, psychologists, neurophysiologists, anatomists, biochemists, professional voice trainers, and endocrinologists have contributed to the study of voice disorders. Multidisciplinary approaches in voice science would be particularly suited for support by the National Research and Training Centers (see chapter 7). These centers should have significant impact on both research and clinical delivery.

Epidemiology

Disorders of voice are increasingly recognized as a major health problem among persons of all ages in all walks of life. The causes of voice problems are diverse and, in many instances, unknown. Despite recognition of the devastating effect of voice problems, there are no reliable data on the prevalence of voice disorders among any postpubescent population. Prevalence data are necessary for adequate planning of health services. Interactions between voice disorders and the environment, vocal usage, and health make it likely that the incidence of disorders is higher in some populations (geriatrics, public speakers, cheerleaders, smokers, alcoholics) than in others. From data on school-aged children, prevalence is estimated to be from 6 to 24 percent. This broad spread is the result of a number of factors, including a lack of a comprehensive classification system, sampling difficulties, training of examiners, and failure to include direct laryngoscopic examinations as part of the screening procedure. For these reasons, prevalence studies are considered to be a high priority. (See also chapter 7, "Data Systems and Population-Based Studies.")

SUMMARY OF RESEARCH RECOMMENDATIONS

Major Scientific Research Opportunities

Laryngeal and Respiratory Physiology

- Evaluate the compartmentalization of the laryngeal muscles with regard to histochemistry and motor unit organization during different gestures, such as inhalation, exhalation, effort closure, phonation, and swallowing.
- Obtain information about the modification of sensory and muscle action feedback to the motoneuron pools during different laryngeal gestures in higher level primates and humans.
- Map the cortical control of the motor and sensory aspects of the larynx.

- Study the regulation of pitch, loudness, register, and quality, with emphasis on the role and interactions of specific muscles and on their aerodynamic properties in relation to changes in the length, shape, configuration, and forces of the vibrating structures. Include vocally trained subjects (singers, actors, public speakers) in addition to normal and impaired subjects.
- Identify the mechanisms of vocal fatigue, and develop theories about the processes of recovery.
- Undertake studies that compare human laryngeal neurophysiology with that of other species proposed as models for human voice production, and explore the validity of study of laryngeal neurophysiology in higher level primates.

Biomechanics of the Larynx

- Investigate the mechanical properties of skin, cartilage, fat, ligament, tendon, and muscle tissues of the larynx.
- Study acoustic and biomechanical source-vocal tract interactions, particularly the effects of vertical movements of the larynx and source filter effects.
- Combine recent developments in the theory of chaos (a new field of study that attempts to find order in seemingly random behaviors) with more traditional linear and nonlinear analysis to describe vocal fold vibration.

Cellular and Molecular Biology and Anatomy

- Acquire new information, which can be developed with existing cellular and molecular biology techniques of immunocytochemistry and in situ hybridization, about the distribution of extracellular matrix molecules, oncogenes, growth factors (for example, epithelial growth factor), and growth-factor receptors, as they relate to normal structure of the larynx.
- Conduct embryology studies of epithelial-mesenchymal tissue interactions that are the driving force behind normal development of the larynx, with attention to how these interactions affect cell differentiation and final organotypic form.

Development, Critical Periods, and Aging

- Characterize voice production as it relates to the developing structure, with emphasis on the regulation of pitch, loudness, register, and vocal quality, with changes in morphological structure.
- Identify the critical periods for optimum development of vocal control, including infancy, puberty, time of auditory impairment, and periods associated with hormonal change, as well as periods when a speaker is recognized as being at risk for developing dysphonias.

- Study the use of physiological feedback and motor learning in training vocal control in the congenitally deaf and in adults who are profoundly deaf.
- Establish the nature of geriatric voice changes, and develop techniques to forestall or prevent them when such changes may interfere with a person's life or livelihood.
- Determine how selective cell death and tissue atrophy affect respiration and voice in aging.
- Collect anatomical, cellular, and molecular information from aged larynxes of a genetically defined species, with emphasis on localization of extracellular matrix components, growth factors, oncogenes, and growth-factor receptors.
- Expand the base of information about the cell biology of joint deterioration in the aging larynx.
- Determine how to improve the secretory process leading to increased pulmonary efficacy.

Major Clinical Research Opportunities

Organic, Idiopathic, and Neurologic Diseases

- Improve the understanding of inflammatory diseases of the upper airway and larynx, including rhinitis, sinusitis, and laryngitis, and develop better therapy.
- Undertake viral typing of laryngeal papillomas and assess their recurrence rate, effects of treatment, effects of cofactors such as smoking and voice abuse, and rates of association with carcinoma of the larynx.
- Evaluate long-term treatment of spasmodic dysphonia and vocal fold tremors with minute injections of botulinum toxin into the intrinsic laryngeal muscles to determine whether this treatment continues to be effective with repeated injections over several years.
- Study the neural control of the larynx to determine why laryngeal functions are particularly vulnerable to neurological disorders causing selective neuronal loss either in the brain stem or in the basal ganglia.
- Clarify the relationships between neuropathology and laryngeal disorders.
- Expand the knowledge about the mechanisms underlying the formation of idiopathic dysphonias, including environmental noise, air pollution, and systemic disease.
- Conduct epidemiological surveys and clarify the precise etiology and pathophysiology, and strategies for prevention, diagnosis, and management of the effects of environmental agents on the upper respiratory tract (nose and sinuses) and the larynx.
- Expand the information about the effects of reduced respiratory capacity on speech and on the neuromuscular mechanisms and reflexes regulating respiratory volume and flow during speech.

Procedures for Diagnosis and Treatment

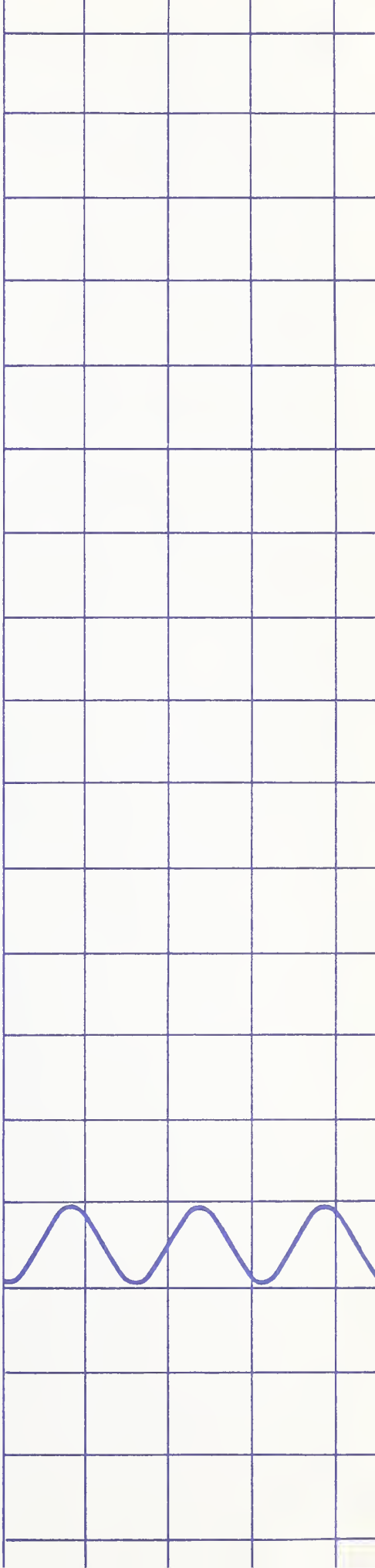
- Expand basic science and clinical information about the pathophysiology, prevention, assessment, and treatment of problems associated with laryngeal surgery and injury.
- Clarify the best strategies for evaluating and treating patients with vocal disorders due to the effect of hormonal and pharmacologic substances on the voice, and find new strategies for prevention.
- Continue systematic investigation of voice production using kinematic, acoustic, aerodynamic, and imaging diagnostic aids to determine which technique (or combination of techniques) is most useful in detecting disorders, documenting changes resulting from treatment, identifying persons at risk for developing voice disorders, setting standards for routine assessment, and improving the understanding of the process of voice production.
- Establish reliable methods for objective voice measurement, including measures of vibratory function, acoustic measures, phonatory function analysis, airflow, improved electroglottography, and repeatable psychoacoustic measures, and identify appropriate tests and develop a convenient, inexpensive equipment armamentarium for performing them so that they may be used by practicing otolaryngologists and speech-language pathologists, and, to some degree, in conservatories and acting schools.
- Obtain further electromyogram data, during different vocal control tasks, on various neurogenic-based dysphonias, and in conjunction with other techniques.
- Conduct human and animal studies of selective intrinsic laryngeal muscle denervation to gain improved understanding of the potential role of laryngeal impairment in many cases of dysphagia.
- Continue development of electronic artificial larynges (external, internal, and implantable), and tracheoesophageal shunts (with and without alloplastic voice prostheses).
- Expand the capability to electrically pace the paralyzed vocal fold to reestablish its breathing function.
- Expand the use of phonosurgical methods through standardized measurements of vocal output before and after surgery, and of simulation modeling to predict vocal outcome before surgical manipulation.
- Establish, through interdisciplinary teams including teachers of singers, the efficacy of various therapeutic techniques for professional and nonprofessional voice users.

Instrumentation and Technological Developments for Diagnosis and Treatment

- Conduct studies of major categories of voice disorders to determine if sufficiently distinctive profiles make it possible to use low-cost acoustic analysis devices for detection and characterization of specific disorders.
- Determine how measures of air pressure and airflow categorize different pathologies, vocal fatigue, test tasks, changes in vocal control strategies, glottal configurations, and other aspects of vocal function.
- Relate observations made from a combination of imaging techniques to each other through appropriate models for use in understanding voice production and application to clinical populations.

4.

Speech and Speech Disorders



CONTENTS

OVERVIEW 131

RECENT ADVANCES 133

GOALS 135

Normal Structure and Function 135

Development and Plasticity 140
Speech in the Aging Population 141

Principal Diseases and Disorders 141

Disorders of Articulation 142
Neurological Disorders of Speech Articulation 142
Neuropharmacological Studies of Speech Disorders 142
Speech in Individuals with Craniofacial Anomalies 143

Speech of the Hearing Impaired 144
Speech Production in Stutterers 146
Swallowing Disorders 147
Research Needs Across Populations 148

Speaker-Listener Interactions 148
Clinical Trials of Intervention Strategies 148
Comorbidity/Co-occurrence Studies 149
Multicultural Aspects of Speech Disorders 149
Genetic Studies 150

Methods and Devices for Analysis and Treatment 150

New Technologies for Research 150
Microcomputer Applications 151
Augmentative Communication Devices 152

SUMMARY OF RESEARCH RECOMMENDATIONS 153

Major Scientific Research Opportunities 153

Normal Anatomy and Physiology 153
Normal Speech Development and Aging 153
Devices for Speech Analysis 153

Major Clinical Research Opportunities 153

Speech of the Hearing Impaired 153
Speech Production in Individuals Who Stutter 154
Speech in Individuals with Craniofacial Anomalies 154
Speech in Neurological Disorders of Communication 154
Research Needs Across Populations 154
Technological Needs Across Populations 154

4.
Speech and Speech Disorders

OVERVIEW

Speech production is one of the most complex motor skills human beings must acquire. To produce speech, air is pushed from the lungs to pass between the vocal folds in the larynx. These folds must be moved into a precise position in the airstream from the lungs so that the mucosal covering is set into vibration by the moving air column. This vibration results in a pulsing of the airflow, which is resonated in the pharynx, mouth, and nose to produce the vowel sounds. For each vowel, the lips, jaw, throat, tongue, and soft palate shape the vocal tract to resonate those portions of the sound spectrum required for listeners to identify that vowel. Most of the information in speech, however, is contained in consonants, which depend upon rapid interruptions in the airflow above the larynx. Some consonants (such as "t") are produced by stopping the flow of air in the mouth and then quickly releasing a burst of air, while others (such as "s") are produced by forcing air through a narrow channel to make a high-frequency noise. Crucial in the production of correctly perceived consonants are the correct location and the speed of movement of lips, tongue, and soft palate from one position to another. The learning required to produce these rapid motions is formidable and requires listening to speech, extracting the acoustic cues that are important, and learning by trial and error how to generate similar sound patterns with the articulators, larynx, and vocal tract.

For conversational speech, the speaker must produce sequences of sounds at a rapid rate. Speech sounds modify each other, so that a particular sound is produced differently, depending upon which sounds precede and follow it. This phenomenon is called coarticulation. It demonstrates the computational complexity required of the central nervous system (CNS) in planning, programming, and executing vocal tract movements for speech. Coarticulatory changes in motor gestures depend not only on the other sounds contained in the syllable but also on sounds contained in syllables preceding and following a sound. Speakers are not aware of these coarticulatory changes, but they contribute substantially to a listener's perception of natural-sounding, intelligible speech.

Slips of the tongue also demonstrate the temporal sequencing required to produce the syllables that make words during speech. Speech sound sequences are "programmed" long before the motor gestures are executed, and sounds can be substituted or misprogrammed among the words in a sentence. For example, "darnboor" instead of "barndoor" is an error in sequencing the

sounds across words when the "d" in "door" is anticipated well before the "b" in "barn" is produced.

There is evidence of the motoric and computational complexity of speech in the frequency with which speech disorders occur following brain injury and in neurological disease. Speech execution disorders, the dysarthrias, occur in almost all diseases of the central nervous system, since any disturbance in movement control will affect the accuracy and speed of articulator movement. Reduced speed and accuracy of movement of the tongue, pharynx, velum, vocal folds, and lips impair speech of patients with Parkinson's disease, amyotrophic lateral sclerosis (Lou Gehrig's disease), Huntington's chorea, and multiple sclerosis, among others. Injury to the brain centers involved in speech learning affect the programming and sequencing of sounds in speech dyspraxia.

Infants spend several years playing with sound and learning to imitate the speech sounds in their environment. Within the first four years of life, the infant must learn to control over 100 different muscles to produce sounds, simply by listening to others' speech. They must simultaneously move the diaphragm, rib cage, larynx, pharynx, tongue, soft palate, and lips in precise ways to make the sounds that they hear. This is a gradual process, requiring many years of practice and learning, with all sounds appearing by 6 years of age. Children refine their speech production for several additional years.

The immensity of the motor learning task that infants must achieve in learning to speak is evidenced by the high frequency of speech production disorders in young children. On entry to first grade, 5 percent of the child population have noticeable speech disorders, such as missing sounds, substituted sounds, or stuttering. The vast majority of these disorders are idiopathic: that is, there is no observable cause for their speech difficulty. Legislation now requires that States provide early intervention for these children to assist them in developing speech normally so that they will not be handicapped throughout early school years. The majority of these speech impaired children do learn normal speech, although a significant proportion of them, about 20 percent, will remain speech impaired to some degree for the rest of their lives.

The importance of normal hearing is obvious for children in learning to identify each sound and in improving their own production of the speech sounds so that others can understand. A child must learn to ignore individual differences among speakers such as pitch, loudness, and speech rate and must attend to the particular acoustic features representing consonants, coarticulatory effects, and vowel sounds. Even the slightest hearing impairment can interfere with speech learning, while profound deafness from birth can prohibit the development of normal intelligible speech. About 0.1 percent of children have a hearing impairment that significantly interferes with the development of normal speech. Hearing aids and many years of speech training from infancy through adulthood are required for children who are born with profound deafness.

Speech development is also disturbed by cognitive disorders such as mental

retardation, and may never be achieved by those with severe mental impairment. Some of these children never learn all speech sounds.

RECENT ADVANCES

Significant advances have been made in the last 10 years in understanding normal speech production and speech disorders. Studies of normal adult speakers have demonstrated the complexities of the speech motor control system. Technological advances have provided instrumentation for measuring lip, jaw, velar, tongue, pharyngeal, and laryngeal movements during speech production. Investigators can now record activity from many of the muscles used to produce these movements.

Such studies have shown that speakers frequently use very different combinations of lip, jaw, tongue, velar, and laryngeal muscles to produce the same speech sound. Different vocal tract shapes can be used by the same speaker from time to time to produce the same sound. Further, no two speakers have exactly the same size and shape of vocal tract, and all use slightly different movements to produce sounds that listeners identify as the same sound. These phenomena are known as motor and perceptual equivalence. For example, when a speaker places a pipe in his mouth, he automatically moves his tongue, lips, and jaw differently to produce the same sound as before, using different articulator movements. He makes this change without any practice. Perceptual equivalence allows two speech waveforms with very different acoustic characteristics to be perceived by listeners as the same speech sound. Perceptual and motor equivalence is evidence for the extensive computational complexity of the human speech perception and production systems. Scientists are attempting to model these systems in an effort to understand them better and to develop prosthetic devices for the speech and hearing impaired.

Scientists have demonstrated in normal speakers that if an error is produced in one articulator, the other articulators, in a fraction of a second, change their movements to compensate. A normal sound results. This change occurs automatically without the speaker's even being aware of the correction being made.

These actions demonstrate the highly sophisticated skills that humans must learn to produce speech. Once speech has been learned, speakers do not continue to rely on hearing their own or others' speech to continue producing normal speech. It is only after many months that an adult speaker who becomes profoundly deaf loses some speech accuracy and requires speech training.

Scientists have demonstrated that once speech is learned, it is highly resistant to disturbances due to a loss of hearing or changes in sensation. Changes in the motor control systems, however, can have devastating consequences. Studies of adults with dysarthria following neurologic diseases and disorders have demonstrated that different regions of the central nervous system control different aspects of speech

production. The subcortical systems, the basal ganglia in particular, have an important role in controlling the speed of speech articulators and speech rhythm. However, the accuracy and speed of articulator movements and the sequencing of speech sounds are disturbed when the cortical regions in the frontal part of the left hemisphere are injured in addition to the basal ganglia.

The detailed neurological organization of the human speech production system is as yet unknown. Studies of monkeys have recently demonstrated that, contrary to limb control, where one side of the brain controls the muscles only on the opposite side of the body, the facial muscles involved in human speech seem to receive input from both sides of the brain. That is, there may be bilateral representation of some of the facial muscles in the brain. Using noninvasive cortical mapping techniques such as transcranial magnetic stimulation, scientists are gaining further information on the cortical representation of the speech musculature in humans.

Significant advances have been made in the study of stutterers. Studies of speech that is produced when a stutterer is fluent have been compared with that of normally fluent speakers and found to differ in subtle ways. The fluent speech of stutterers, which sounds normal to listeners, is slower and is less able to compensate for errors during speech. This finding indicates that there is some abnormality in the speech motor control system that stutterers must overcome when they produce fluent speech. Scientists have recently found neurophysiological differences in brain function that may interfere with speech production in adult stutterers.

Behavioral geneticists have shown that pedigree analyses of speech disorders in the families of stutterers demonstrate significant family histories in a number of chronic adult stutterers. This finding suggests a genetic predisposition for stuttering in some families.

New help has recently become available for the most severely speech-impaired persons, those "locked in" from birth with cerebral palsy, or who are made speechless by head injury, laryngectomy, or neurological diseases such as amyotrophic lateral sclerosis (ALS). Talking machines, using microcomputers, are being used by cerebral palsied children to produce speech phrases at the touch of a switch. These children can learn new words or speech sounds using keyboard entries. Dozens of new devices have become available in the last decade allowing previously locked-in patients to converse, use the telephone, and answer in the classroom.

Recently developed microcomputers can automatically analyze speech in real time and produce a visual display on a computer screen while a speaker is saying words and sentences. These computer systems are now being used to train deaf children to speak. Hours of speech training have become possible using relatively inexpensive microcomputers, which these children enjoy using. This advance allows deaf children to work on speech training outside of school hours so that they can participate during regular school hours in classes with normally hearing children.

New technological advances have also opened significant new avenues for the study of vocal tract movements during speech, chewing, and swallowing. An existing national center for speech research allows scientists to study movements inside

the mouth. Such studies were previously impossible without the use of dangerous x-rays. Movements of the tongue, the major speech articulator used also for chewing and swallowing, can be measured by employing miniature pellets that are traced by a machine and displayed on a computer screen in a reconstruction of the inside of the mouth. Scientists from around the country can now travel to this center to conduct speech studies.

Other techniques such as miniature magnetometers and ultrasound are also being used to study tongue, pharyngeal, and velar movements during speech, chewing, and swallowing. Such technologies have made studies of tongue movement feasible and have decreased the amount of time and effort required to study speech. With computerized signal processing, the analysis of movements, muscle actions, and speech waveforms is now feasible in both the clinic and laboratory. With the new technology, the understanding of normal speech and of speech impairments of all types will expand dramatically in the next decade.

GOALS

Normal Structure and Function

The overall properties of the speech waveform depend on the structures of the upper vocal tract. Because of the hidden nature of the important structures for speech articulation, investigators are quite poorly informed about them.

Modern imaging techniques (see below) should allow great progress in understanding normal variation, particularly with respect to soft tissue. At present, more is known about the variations in bony structures than about those in soft tissue. Cephalometric studies and other classifications of bony tissue by dental scientists and physical anthropologists have led to extensive knowledge of the variations in size and shape of the bones of the head and neck. These structures can be effectively studied postmortem. The study of soft tissue, however, requires direct visualization in the living human, which is a more difficult task.

A particular need exists for measures that will enable visualization of the whole vocal tract in three dimensions. In producing the speech acoustic wave, the upper vocal tract acts as a filter on the output of the larynx, and the properties of the output acoustic wave can be predicted by using computer simulation analysis.

To synthesize the acoustic output of the vocal tract for an isolated, steady state vowel, the cross-sectional area of the vocal tract from larynx to lips must be known section by section. Since most of the current information on tract shapes comes from lateral view cineradiography (x-ray motion pictures), the third dimension is not well estimated in present simulation models. More needs to be known about variation in the third dimension.

Knowledge about the region of the pharynx and velopharyngeal port is particularly deficient. Conventional study

of phonetics has focused on a view of the activity of the tongue in which the position of the tongue front is overemphasized. These misunderstandings are in need of correction, both to understand normal function and also to assess the effects of alteration, by surgical means, of the gross deviations in regional morphology in craniofacial anomalies.

Studies of vocal tract morphology, with computer modeling, should have important applications in predicting the effects of vocal-tract tissue removal for cancer surgery or for correction of congenital anomalous structure.

- Population-based studies of gross structure of the upper vocal tract, especially of soft tissue, are needed. These studies have important applications in assessing and predicting the effects of therapeutic or traumatic change. Recent developments in imaging techniques make such studies feasible in the immediate future.

While substantial advances have been made in the last decade in understanding the biochemical and mechanical properties of striated muscle, the orofacial system has received limited attention. For most of the limb muscles, three or four types of fiber are commonly found, and data are available on their properties. The distribution of fiber types of the muscles of the jaw, tongue, lips, and larynx, and their changes in ontogeny and phylogeny, are less well understood. There is some indication that muscle tissue in the parts of the body innervated by cranial nerves is different from that innervated by cervical and other spinal nerves. Because the conventional histochemical procedures utilized for analyses of limb muscle tissue do not yield the same reactions in orofacial muscles, there is a question about the suitability of these techniques for study of orofacial muscles. For this and other reasons, it seems essential to explore tissue properties by immunochemical and ultrastructural techniques. The ultrastructural studies that have been crucial in determining the basic characteristics of contraction and control of the muscles of special function, such as those of eye movement, have not been conducted for upper airway muscles.

Data on properties of muscle tissue are essential in understanding normal differences in speed of contraction of the different muscles. Likewise, traumatic or surgical reinnervation of the orofacial muscles cannot be evaluated without knowledge of basic muscle tissue properties. For example, there is no explanation for the report that facial muscles, upon biopsy, retain their biochemical viability, while limb muscles die after denervation. Presumably, more complete evaluation of tissue properties (with the appropriate accompanying muscle fatigue studies) will allow scientists to understand the apparent extraordinary histochemical and electrophysiological resistance of orofacial muscles to fatigue.

- Analyses of orofacial muscle tissue characteristics, utilizing conventional and newly developed histochemical, immunochemical, and ultrastructural techniques will allow investigators to understand their contraction patterns and biomechanical behavior, and to interpret the changes associated with many disease states.

The acquisition of better knowledge of properties of orofacial muscle tissue must be accompanied by research directed toward specification of the motor innervation of these muscles. The location, boundaries, and organization of lower brain stem motor nuclei reported in textbooks is based, almost exclusively, on observations using now questionable nerve lesion-degeneration methods. Such analyses, as argued in many recent neuroanatomical papers, will need to be undertaken again, using modern axoplasmic tract tracing techniques. While these procedures have been available for more than 10 years, studies of cranial motor nuclei have usually been performed with nonprimates, and available information is limited to the motor neurons innervating the facial and jaw muscles in such species. The organization of the canine hypoglossal nucleus has been studied recently, and critical connections between it and other centers have been revealed, but the lower brain stem innervation of the velopharyngeal muscles remains poorly understood.

In addition to determining the lower brain stem subnuclear organization for various orofacial and laryngeal muscles, quantitative analyses of the motor neuron pools are needed. Particular areas of promise include determining the size, numbers, and density of the motor neurons in relation to the muscles they innervate. Recent work, although limited, indicates, for example, that the muscles of the perioral region are innervated by significantly larger motor neurons than those innervating periocular or frontal muscles. There is almost no information, however, on the relative sizes of cranial motor neurons in the hypoglossal, trigeminal, glossopharyngeal, and ambiguous nuclei. Such data would allow prediction of the relative conduction speeds of the various associated motor neurons and also an assessment of the relative distributions of alpha and gamma motor neurons.

During the past decade, there has been a substantial change of direction in biochemical studies of the central nervous system. In the area of speech, there is great need to examine the biochemistry of lower brain stem motor nuclei. The results should allow an evaluation of the range of neural transmitters and neuromodulators in this region and an ability to predict the influence of pharmacologic agents on orofacial function. A better understanding of the biochemistry might enable a better understanding of regional systemic disease processes. ALS patients, for example, seem to show their earliest impairment in tongue muscle function. It may be that this pattern is related to the apparent, but largely unstudied, biochemical uniqueness of the hypoglossal nucleus.

- There is need for further specification of the motor innervation of the orofacial muscles and their associated lower brain stem motor nuclei. Analysis of the size, organization, and biochemistry of the brain stem nuclei will provide a basis for interpreting neuropathology associated with disease and deviant brain stem function.

Sophisticated dynamic perturbation techniques, adapted from studies of limb musculature, have been applied to the muscles of articulation. These studies, taken overall, lead to the conclusion that

even the most rapid speech movements are subject to sensory correction. Information is lacking, however, on the receptors involved in these control processes, their afferent projections, and the way in which their afferent information is processed in the various relevant brain centers to shape and correct motor output.

The kinds of receptors involved in transducing motor output information need to be determined. As with other aspects of the orofacial system, there is a lack of similarity to limb structures. The characteristics and distribution of these receptors appear to be different.

Some recent human studies, utilizing conventional psychophysical techniques, offer new information on the accessible receptor sites on the facial skin. Microneurography, a technique in which microelectrodes are placed in sensory nerve fascicles to record single afferent discharge patterns from human mechanoreceptors, complements psychophysical techniques. Using the techniques together, investigators expect to establish the distribution of types of receptors, their location on the skin or mucosa, the receptive field sizes, shapes, and sensitivity, and information on patterns of discharge with movement.

While detailed studies of the neurophysiology of sensory motor interaction are important, temporal details of the behavioral interaction of sensory and motor input in determining speech behavior are needed. For example, perturbation studies can tell something about the way articulators are linked in the temporal plan for an utterance. These studies should be important in comparisons of normal populations with groups such as stutterers, who are believed to have defects in planning in some ill-specified aspect of sensory motor integration.

- Electrophysiological, morphological, and psychophysical analyses of sensory mechanisms associated with orofacial sites are needed, given the evidence accumulating over the last decade that sensory influence plays an important role in orofacial motor control, both in the planning stages before initiation of movement and in the moment-to-moment shaping of motor output. Some disorders of speech may be associated with deficits in processing sensory inputs.

The role of CNS centers in control of speech production is presently assessed largely by clinical studies that document the effects on speech behavior of damage to such centers of the motor and sensory strips of the cerebral cortex, the cerebellum, and the basal ganglia.

During the past 10 years, attempts have been made to document the role of these various centers in the control of phonation in primates and also in the control of the upper vocal tract components of chewing and swallowing. However, there is no nonhuman primate activity directly analogous to speech. Investigators are therefore dependent on such noninvasive techniques as positron emission tomography (PET) for the determination of the CNS centers active in various kinds of speech acts.

Purely anatomical studies are important, both for revealing the CNS interconnections important in speech motor control and for explicating the differences in control

pathways between vocal tract and limbs. For example, while the deep cerebellar dentate nucleus for the limbs projects, via the thalamus, to the primary motor cortex, the orofacial dentate nucleus projects to the nonprimary motor cortex. These differences in pattern of projection must reflect in a specific way the differences in the pattern of control of limb orofacial versus musculature.

- Modern neurophysiological neural-tract tracing and imaging techniques may be expected to contribute to the understanding of the way in which different CNS centers contribute to speech motor control.

While the function of the muscles of the head and neck are described in many textbooks, these descriptions are based on anatomically plausible actions. The way that muscles function in vivo, and the degree of plasticity in function that individuals can use to attain a given vocal tract movement, are at present the subject of active research by a number of groups, who in the last few years have developed the capability for electomyographic research in humans.

A recent study using cross-correlational analysis, for example, has shown that the muscle synergies of the mandibular system are different for speech and chewing tasks, perhaps because of specifiable differences among task demands. Further studies for all the upper vocal tract muscles are needed to specify the correlation patterns among muscle patterns and their cross-task changes for chewing, swallowing, and arbitrary execution of the full range of movement of the articulators. Such information will be useful in deciding on strategies for reconstruction after surgery.

- Research on the full range of upper vocal tract activities that specify the structure muscle-function relationship in speech, chewing, swallowing, and arbitrary voluntary articulator motion is needed.

A complete model of the behavior of the upper vocal tract has several components. First, information is needed about the temporal sequencing of the articulators for the production of a given phonological message, and how this temporal sequencing changes with changes in stress or rate of speaking. Second, information, constrained by equations of motion, is needed about the shapes that articulatory trajectories can assume. Third, a model of the articulators in the vocal tract, which can be used to synthesize speech, is needed.

Articulatory models have been developing in the last decade. The development of means of visualizing the movement of the articulators, such as the x-ray microbeam, has resulted in systematic investigations of articulatory behavior. Empirical investigations of speech and nonspeech behavior have generated models of articulatory trajectories with specifiable parameters. All of these types of investigations should continue.

- Empirical data from investigations of articulator movements should be used to develop articulatory synthesis. The synthetic acoustic output can then be compared with sample acoustic productions.

Development and Plasticity

In many significant ways, the infant lays hold of the environment with the mouth. Besides its obvious importance in receiving nutrition, the mouth of the infant is a means of exploring and learning. The mouth is also the means of sound production, and even deaf infants have been observed to engage in cooing and babbling. How these early sounds may lay the foundation for development of speech and language is not well understood, but recent evidence converges on a principle of continuity; that is, the sound patterns of infancy are consistent with, and may lead to, the sound patterns used by the young language learner.

Many communication disorders appear during childhood or are thought to be rooted in developmental processes. Therefore, to understand these disorders—and to treat and prevent them—it is essential that the processes underlying communication development also be understood. Developmental changes are extensive and probably interactive. During infancy and early childhood, the three major systems of speech production—respiratory, phonatory, and articulatory—undergo major changes in anatomy and function. As an infant is exposed to language, the infant begins to produce more and more sounds like those in the parent language, even though it is physically impossible for the infant to produce exact physical replicas of adult speech sounds. The refinement of speech production extends well past the time that the child enters school, but appears to be essentially mature by the onset of puberty.

Many authorities believe that a child's capacity to learn the sounds of a language is markedly diminished after the age of puberty. This thinking has led to the concept of *critical* or *sensitive* periods in speech and language development. Factors that interfere with accomplishments during critical or sensitive periods may place a child at risk for communication disorders. Conceivably, more than one critical or sensitive period may exist. Whatever the number, research is needed to identify the ages and developmental correlates of these periods. This issue is particularly important in regard to deafness and hearing impairment. It is frequently remarked that children who lose their hearing before they have learned to talk (the prelingually deaf) face much greater obstacles in oral communication than children who lose their hearing after they have acquired speech (the postlingually deaf).

Although data on the development of speech and language have grown substantially over the last decade, an adequate understanding of this process requires large-scale research programs—research on special populations considered to be at risk for communication disorders, and the continued development of tools and techniques suited to the study of infants' and children's sound production. Special priority should be given to the following areas of research.

- There is serious need for both retrospective and prospective studies of development related to risk for later delays in speech acquisition. From these studies, it should be possible to predict communication disorders from developmental precursors.

- Large-scale studies of the comparative development of speech and language behaviors in normal and abnormal population are required to give an accurate picture of the ways in which communication abilities develop in children. There is a special need to evaluate the concept of critical periods.
- Continued efforts should be made to refine existing techniques for developmental research and to create new tools for research in this area. It is particularly important that suitable methods be developed for acoustic and physiologic studies of children's speech. These methods are important not only for research purposes but also for clinical applications in assessment and management.

Speech in the Aging Population

There are age-related changes in respiration, phonation, and upper airway functions, all of which are related to speech. It remains to be determined the extent to which specific senescent changes impair speech.

Findings of some investigators suggest that the speaking rate may be slow overall in a geriatric population with no overt signs of specific pathology. There is also some indication that articulator movements may be somewhat more extreme, perhaps because of slight continuing enlargement of the vocal tract with age. However, there are as yet no large-scale, chronologically detailed studies of the effects of aging on either the acoustics or the kinematics of speech.

Such studies will be important at a practical level to provide control data for comparison with populations with such disorders as Parkinson's disease, which typically affect older groups.

- There should be studies of speech production by normal adults, grouped by age, to serve as control data for assessment of the speech effects of the dysarthrias and other disorders that affect older populations.

Principal Diseases and Disorders

Disorders of speech cover a wide variety of problems but essentially relate to difficulties in producing the sounds, syllables, and words of spoken language. These disorders may occur as a consequence of hearing impairment, neurological deficits, congenital defects, trauma, pharmacological agents, and interference with typical development. In many cases, however, the precise cause is unknown.

Speech disorders severely handicap affected individuals. They limit the ability to function effectively in a world in which communication by speech is a major determinant of economic, academic, and social achievement and status. Thus, it has become increasingly important that diagnosis, prevention, and treatment (including rehabilitation) be improved.

A basic understanding of these disorders may lead, among other benefits, to the development of therapeutic strategies that can improve performance in ways that

can be taught, although they cannot be learned spontaneously.

Disorders of Articulation

A major class of speech disorders is a group whose cause is idiopathic (unknown) as opposed to those of known origin (for instance, due to neurological deficit). According to data gathered by the American Speech and Hearing Association, 40 percent of clients seen by school speech clinicians have such disorders of articulation, and 99 percent of clinicians see such children. Studies that group children with idiopathic articulation disorders on the basis of different speech profiles should be continued. Such groupings may lead to identification of different precipitating factors, which also may lead to improved preventive or therapeutic procedures. Knowledge is needed of which aspects of idiopathic deficiencies of articulation disorders are due to a deficiency of the child's linguistic system (knowledge of the sounds conveying the message) and which are due to the child's motor system (ability to make the desired sounds). This study would allow therapeutic approaches to capitalize on the most appropriate strategy.

There has been interest in the last few years in a group of severely articulatory deficient children who are labeled as having "developmental apraxia of speech." If a discrete group of children does exist fitting the symptom description for apraxia of speech, the assertion that they are less responsive to traditional therapies than other children needs to be verified, and also whether there are more effective procedures to be used with them.

- An ability is needed to identify appropriately and in a timely manner those children at risk for long-term problems of articulation of various types.

Neurological Disorders of Speech Articulation

At present, different laboratories produce a variety of measurements of speech articulation for the neurologically impaired. A better understanding is needed of the effects of the various types and amounts of motor impairment on speech. Such understanding could guide decisions about what aspects of speech should be measured in these patients and lead to the construction of standardized assessment profiles based on perceptual, acoustical, and physiological data. Standardized profiles would allow information to be pooled from patients at many different laboratories and clinics.

- Standardized assessment profiles should be developed for neurological disorders, based on perceptual, acoustical, and physiological data, which taken together will provide a firm basis for understanding neurologically impaired functions of respiration, phonation, and articulation.

Neuropharmacological Studies of Speech Disorders

Speech is often affected in diseases altering neurotransmitters in the brain, such as following a loss of dopamine in the

substantia nigra in Parkinson's disease. Further, neuropharmacological treatments can have profound effects on speech production.

- Careful studies are needed in patients with disorders affecting selective neurotransmitter systems, to determine which types of transmitter systems are involved in the speech motor control.

Speech in Individuals with Craniofacial Anomalies

Perhaps the most significant new direction in research on cleft palate and craniofacial disorders is the recognition that individuals with orofacial clefts are etiologically heterogenous. This recognition is a relatively recent development, coinciding with the growth of the fields of clinical and molecular genetics.

In 1970, research data indicated that only 3 percent of individuals with clefts of the lip and/or palate had associated anomalies or syndromes. More recent studies, published in the 1980s with the expansion of clinical genetics studies, have shown that 50 percent or more of children with clefts have associated abnormalities or syndromes, and over 400 such syndromes have been described. To date, however, little information is available to indicate the relationship between communicative impairments and multiple anomaly syndromes, which include cleft palate. Two questions must be asked. First, do specific syndromes of clefting have predictable patterns of communicative impairment, including disorders of articulation, language, resonance, and hearing? Second, can an assessment of communication disorders assist clinicians in delineating and diagnosing multiple anomaly syndromes? The pursuit of the answers to these questions will require close working relationships among speech-language pathologists, audiologists, and geneticists (both clinical and molecular). The addition of communicative symptomatology to the already well-defined features of recognized syndromes will expand the phenotypic spectrum (clinical picture), natural history, and prognosis, which will ultimately result in better patient care.

The same approach can be applied to many other genetic syndromes. There are several hundred syndromes that have various CNS disorders as features, with one or more types of communication disorder. Some of these syndromes occur frequently in the general population, with hundreds of thousands of individuals affected. These include neurofibromatosis (one of the most common genetic diseases) and many of the muscular dystrophies, including myotonic dystrophy (Steinert syndrome), facio-scapula-humeral muscular dystrophy, and Duschennes muscular dystrophy. In some of these disorders, the communicative impairment may be the first noticed symptom because of the absence or minor nature of other physical symptoms.

- Studies of the clinical history of patients with orofacial anomalies are needed, which differ from conventional studies in two notable respects: (1) all aspects of the presenting syndrome must be assessed, and (2) the full range of communication defects must be assessed, both before and after treatment.

Speech of the Hearing Impaired

In the last decade, a general consensus has developed on a strategy for intervention following prelingual hearing loss. Children who lose hearing prelingually can be classified in groups depending on the degree of loss. In general, speech is not audible to children whose loss is greater than 60 dB; such children require hearing aids and speech intervention as well. A group with hearing losses from 90 to 120 dB cannot hear conversational speech with good modern aids, but can receive significant information about speech that will lead to speech improvement under proper therapy. Finally, children with losses greater than 120 dB can learn speech only through nonauditory means.

Within the past 20 years or so, virtually all children in minimally supportive environments have begun to receive the appropriate amplification to make maximal use of their residual hearing. Children in schools for the deaf, then, are increasingly concentrated in the "profound" and "total" categories, or have multiple impairments. Most recently, children in the profound and total categories have often been provided with cochlear implants, which are likely to bring information useful in developing speech. Tactile aids have also been provided to children in these categories.

Research should be focused on needs associated with the development of speech, particularly in those groups with either newly acquired hearing (through cochlear implant or modern hearing aid) or prelingually very limited hearing.

A first question is the assessment of the nature of speech production in the members of these groups who produce at least moderately intelligible speech. It has been claimed that there is a common pattern of errors in prelingually deaf, orally trained individuals with various degrees of residual hearing and intelligibility. This finding implies either that training practices are fairly uniform, that the nature of the speech produced is insensitive to the fine-grained details of the training and the amount of residual hearing, or that the analysis used was too insensitive to detect differences in pattern. For further training, it is important to distinguish among these possibilities.

- Careful studies are needed of the articulatory characteristics of severely impaired speakers, with a detailed accounting of the risk factors such as educational history and residual hearing.

Studies are needed that use more refined measures of speech production in these groups. Direct studies of articulation are also needed, since only in this manner can investigators distinguish between, for example, faulty management of the aerodynamic characteristics of speech and faulty articulator placement. For example, fricative sounds are poorly made in this group. However, it is not possible to tell by acoustic analysis alone whether a given child makes "s" poorly because the child fails to take in sufficient air, fails to abduct the vocal folds with appropriate timing to build up pressure behind an oral occlusion, or fails to direct the air jet by placing and shaping the tongue tip to direct the air jet across the teeth.

A closely related need is the study of the factors that make the speech of the hearing impaired unintelligible, using modern analyses by synthesis techniques. Several attempts of this kind have been made in the past decade, by using acoustic techniques to manipulate the acoustic speech signal to specify the changes that would have the greatest effects on intelligibility. These studies can be extended by using articulatory data, using an articulatory synthesizer, to investigate the separable articulatory factors for their individual effects. Thus, it is possible to synthetically separate the effects of abnormal use of different parts of the system on intelligibility.

- With the combined development of dynamic models of articulation in normal speakers and the acquisition of detailed articulatory information in hearing impaired speakers, the characteristics of articulation that are most important in causing the deterioration of intelligibility should be investigated.

Ingenuity has been used in the development of training protocols that are sufficiently explicit to give guidance to teachers in schools and also in the development of more computer-based training aids to relieve teachers of some training burden. This effort must continue.

- On the basis of information on the articulatory characteristics of the speech of the hearing impaired, training aids and protocols should be developed that will deliver articulatory information in a focused manner and assess progress in learning independently for different components of speech production.

Discussions of speech training for the hearing impaired have focused on children with substantial losses, sustained prelingually. There is an assumption in the literature that there is a critical period for loss, focused some time in the speech-learning years from one to three. This assumption has never been tested by a large-scale, careful study of the effects of different degrees of loss at different points in the life cycle. This is a theoretically interesting question, as well as one with substantial practical consequences.

While it is well known to clinicians and is discussed in textbooks on rehabilitation that substantial hearing loss at any age is likely to be followed by deterioration of speech, which may be prevented by the appropriate provision of clinical training-support services, very little is known about the form that the deterioration takes and about the forms of speech training that are effective. Such information is particularly needed with the increase of an aging population likely to develop substantial losses and also the increase of mainstreamed children who may have progressive losses and need a focused source of speech training.

- There is need for large-scale studies of the effects of hearing loss on speech at various times in the life cycle. These studies should yield explicit information on the articulatory details of the resulting speech. Furthermore, there should be comparative studies of remediation protocols for groups who sustain loss postlingually or move into a hearing environment by mainstreaming.

Speech Production in Stutterers

Stuttering is a disorder that has a relatively high prevalence. Different studies estimate the prevalence of stuttering in school-aged children to be between about 0.5 percent and 2.2 percent of the total population. Most of these children receive some kind of intervention. In spite of the high prevalence of stuttering, the large amount of intervention attempted, and a general lack of understanding of many basic aspects of the problem, very little research in stuttering is presently funded. No active research grants from the NIH have stuttering as a primary focus, and only three current grants include research on stuttering. This is an area in which training should be supported. The effort would lead to an increased number of proposals for research grants and attract new investigators to the area.

It now appears, after nearly a decade of intensive research, that stutterers' use of the speech production mechanism during their typical or fluent speech is grossly similar to that of their normally fluent peers. Stutterers do exhibit, however, subtle aberrations during their perceptually fluent speech production. While it is now known that laryngeal disruptions, once thought to be central to the problem, do occur during stuttered speech, it has become clearer that much more attention should be paid to stutterers' abilities to produce the complex temporal/spatial coordinations among respiration, phonation, and articulation necessary for fluent speaking.

Research in the area of stuttering can be separated into needs to assess predisposing or precipitating *causes* versus needs to assess perpetuating or aggravating *factors*. Once the problem has begun, for whatever reason, there is a variety of factors that maintain the problem, and both aspects—the predisposing and perpetuating factors—need to be better understood to improve diagnosis and treatment.

With regard to precipitating causes, there is a clear need to assess stutterers' speech motor control for fluent speech to assess whether the difficulties that they experience during speaking are specific to instances of stuttering or are pervasive throughout the entirety of their speech. In addition, the need to define and measure temporal/spatial coordinations among respiration, phonation, and articulation during fluency continues. These speaking abilities need to be correlated with current technology for assessing central processes (for example, brain function) to determine further the role that central processes may play in descriptions of stutterers' speech production.

Research into the genetic aspects of stuttering, through such mechanisms as twin and family studies, is needed to clarify the extent to which heredity may predispose individuals to stutter. It is, however, urged that the complex interactions between nature (genetics) and nurture (environment) be taken into consideration through appropriate genetic as well as statistical modeling.

Perpetuating factors, while not causal in any primary sense, clearly have the potential for aggravating or maintaining a stuttering problem. There is a need for studying

the communicative and related interaction between stutterer and listener, particularly mother and young stutterer. Related to these interactions is the notion of critical periods, a period of time when a child's speech fluency might be particularly vulnerable to external or environmental stress. These critical periods need to be studied in boys and in girls. The results may give some clues as to why boys manifest the problem more than girls.

There has been a continued interest in subgroups. Once again, it may help to separate subgroups etiologically and behaviorally. At present, it is not known whether behavioral differences are correlated with etiological differences. Stutterers with different numbers and types of disfluent behavior may have different causes, and these differences may influence their treatment and recovery.

There should be some careful prestuttering versus poststuttering therapy studies. Therapists need to know what changes in therapy are most related to stuttering and which of them may be predictive of long-term change. While some of these measures may be acoustic, perceptual, or physiologic, such psychosocial issues as locus of control, codependency, and attitudes towards communication should also be considered. Likewise, there is a need for careful, objective, comparative studies of various types of therapy.

Results of research of the past 10 years make it clear that stuttering does not exist in a vacuum. Stutterers, particularly children, also frequently exhibit phonological, language, and voice problems and their recovery from stuttering may or may not be influenced by these concomitant difficulties. There is a great need for systematic study of the number and nature of these concomitant problems, how they may influence a stutterer's recovery, and what they may say about the etiology of stuttering and about the relationship between fluency, sound production, and language.

It is expected that refinements in neuropsychological methodology will help investigators better understand the role of cognition, perception, and neuromotor functioning in stuttering. Advances in the area of brain imaging should also be applied to stuttering, for improving the understanding of structural as well as functional similarities between stutterers and normally fluent speakers. Research is needed to assess the degree to which this information improves clinicians' abilities for differential diagnosis and hence decisions regarding treatment.

- Studies of stuttering are needed that are focused on (1) measures of coordination among laryngeal, respiratory, and upper articulator coordination, (2) the possible existence of subgroups of stutterers with different symptom constellations and family histories, (3) influence on stuttering of communicative and related interactions between stutterer and listener, and (4) the effects of standard therapeutic regimens on various aspects of stutterers' articulatory behavior.

Swallowing Disorders

Significant swallowing disorders, the dysphagias, accompany most injuries or

diseases affecting the central nervous system. However, knowledge about the motor control system is extremely limited for both the reflexive and volitional components of swallowing. With improved measurement techniques for analyzing swallowing disorders and the recent advances in brain imaging techniques, the potential for gaining significant new understanding of the human swallowing mechanism is great. Studies are also needed to understand better each of the muscle actions during swallowing and how they are affected in various structural and neurological disorders. Since the human vocal tract is structured very differently from that of animals, animal studies may not be relevant.

Research Needs Across Populations

Speaker-Listener Interactions

Research is needed to assess a wide variety of verbal interactions and nonverbal interactions (facial gestures) between speakers and listeners. It is presently unclear how these interactions may influence the development and nature of a speaker's oral communication, particularly in speech disorders. More information is needed regarding the important communicative and related interactions between mothers and their young children who are developing oral speech and how these interactions may influence positively, neutrally, or negatively the onset and development of various speech disorders: for example, speech misarticulations, stuttering, or speech disruptions due to craniofacial anomalies. Without this information, it is quite difficult to meaningfully counsel parents of children with speech disorders about what to do in the home environment to facilitate or help change the children's speaking behavior. Such research would also assist clinicians to assess those facets of speaker-listener interactions associated with particular speech disorders and plan intervention strategies accordingly.

Clinical Trials of Intervention Strategies

Clinical trials are needed to evaluate systematically a variety of intervention strategies used in the remediation of various speech disorders. The number and nature of these strategies is such that there is a need to establish some guiding principles by which to derive new therapeutic regimens as well as improve those in existence. While a variety of procedures seem to work for various speech disorders, in many cases it is unclear how and why they work. Developing such understanding is crucial to the standardization of these procedures across a wide number of clinical settings. Importantly related to these clinical trials is the need to study empirically the reasons for the occurrence of relapse *after* apparently effective therapy, as well as procedures that would minimize the occurrence of relapse. Indeed, with some speech disorders (for example, stuttering), achieving an initial change in the disorder is not as difficult as maintaining that change. Systematic, empirical study needs to be focused on the reasons why relapse takes place.

Comorbidity/Co-occurrence Studies

Research is needed to study the nature and number of problems that occur together in one individual with a speech disorder: for example, speech sound misarticulations exhibited by stutterers and conductive hearing loss in persons with a cleft palate and, most particularly, speech and language problems. Interactions among these problems are such that they often significantly impede progress in therapy. For example, strategies to change misarticulations in a stutterer may actually worsen the person's stuttering. These co-occurring or concomitant problems may suggest different etiologies and hence have a significant impact on the type, length, and eventual outcome of therapeutic intervention. At present, however, little is known about the prevalence and nature of comorbidity for speech disorders and how comorbidity influences the evaluation, diagnostic labeling, and treatment for the co-occurring problems.

Multicultural Aspects of Speech Disorders

While various studies have been conducted to provide epidemiological data on various disorders of speech in the general population, only scant reference has been made to racial and ethnic minority groups. Basic epidemiologic information is needed on the prevalence and risk of occurrence of speech disorders in multicultural populations. More research is needed in the area of nondiscriminatory assessment. Currently, there are few appropriate instruments for the assessment of the speech disorders of linguistic minority populations. Similarly, there is a need for more normative data on the development of phonology and grammar in multicultural populations.

Multicultural research is needed in the area of organically based communication disorders. Because of gaps in existing research data bases relative to multicultural populations, accurate information is severely limited. For example, the incidence of cleft lip or palate is known to vary cross-racially, with the highest incidence in the Asian population, mid-incidence in the white population, and lowest incidence in the black population. However, risk figures, which are commonly used to predict the probability of recurrence of cleft lip or palate within a given family, were derived from epidemiologic studies of white populations. No data on risk of recurrence currently exist for multicultural populations. Multicultural research also is needed in the area of fluency disorders. There is evidence of sociocultural influences on the development and maintenance of disfluent speech or stuttering. In general, the paucity of research on communication disorders among black, Hispanic, Asian, and American Indian populations within the United States represents a serious void in the knowledge base, which must be addressed immediately, especially in view of the demographic trends projected for the next two decades when one of every three persons in this country will be nonwhite.

Genetic Studies

The need for genetic studies has been mentioned at several points in the foregoing discussion. The Task Force is aware that there is a general need for increased attention to this area for all speech disorders.

Methods and Devices for Analysis and Treatment

Technology should be central to many of the scientific and clinical concerns of the NIDCD. It may be applied to dynamic and static visualization of the speech production process, including soft tissue anatomy; respiratory data acquisition; clinical training and assessment of function; assistive and prosthetic devices; and objective evaluation of the effects of treatment. A primary focus of technological development needs to be in the area of instrumentation to measure and evaluate the oral and laryngeal structures. Orofacial behaviors for speech and swallowing are especially difficult to visualize because they are hidden from view, and any instruments placed within the cavity tend to interfere with normal movement.

New Technologies for Research

The oral mechanism involved in the production of speech and swallowing involves a very large number of structures that interact in complex ways. The examination of any particular piece of this puzzle is made more difficult by the fact that all activity takes place in the hidden recesses of the oral, nasal, and pharyngeal cavities. Thus, attempts to directly measure speech production are easily confounded by the distortion of the speech event that occurs when the measuring instrument is inserted into the region.

The most successful measures of speech production are unobtrusive instruments. The first type of unobtrusive measurement is indirect, such as (1) examination of the acoustic spectrum and inference of vocal tract shapes, and (2) measurement of airflow and pressure, and aerodynamic modeling of speech flow and vocal tract movements. The second type is measurement in which instruments inserted into the oral and nasal cavity have only minimal effect on speech movements. These instruments are valuable because they directly measure articulator movements. They include point tracking systems, which examine articulator movement over time, such as LED tracking systems, electromagnetic inductance, x-ray microbeam, and strain gauges. Two other direct and minimally biasing measures are electromyography (EMG), which provides unique muscle contraction information, and dynamic electropalatography, which similarly provides unique information about tongue-palate interaction.

The last category of instrumentation involves noninvasive instruments that produce direct measures of articulation but insert nothing into the oral/nasal area. These are primarily imaging techniques, which use waveforms such as x-rays, sound waves (ultrasound) or magnetic fields (MRI), and radio waves to image and distinguish between soft tissues in the body.

Use of each of these types of instruments adds to the knowledge of speech

function in different ways. For example, minimally invasive direct measures are rapid enough to provide essential speech timing information. This capability is critical, since the timing of articulator movement is one of the keys to speech organization and motor control. However, minimally invasive techniques generally measure only one portion of the vocal tract, such as lip and jaw movement, so that only a small part of the event is described. The opposite situation occurs with noninvasive imaging techniques. These measures capture the entire event, providing for the first time a holistic view of tongue and vocal tract movement. Some imaging techniques, however, are presently too slow to provide information about the split-second timing used in continuous speech. Thus, each technique needs to be enhanced to overcome its difficulties. Equally valuable is the combination of techniques to define the synergies among timing, shape, and the acoustic output of vocal tract movement.

An important aspect of instrumentation development is its application to the study of disorders. Basic instrumentation needs to be applied to normal and clinical populations to create data bases that researchers and clinicians can tap into. For example, swallowing has recently emerged as an area of interest to and need in speech research. Very few techniques have been applied to this research area, yet many disorders affect swallowing. Furthermore, infant oral development is intricately related to changes in swallowing pattern.

A final and vital area of research is the extension of instrumentation used in speech research. This includes development, refinement, and application of instrumentation. Technology currently used in other medical and engineering settings should be pursued with respect to its use in speech and swallowing research. The instruments currently being used should be improved regarding sampling rate and accuracy of event reproduction. In addition, current instrumentation should be made more available in the research area, especially in joint efforts among researchers and research groups, particularly when each researcher brings unique talents to the venture.

- Particular emphasis should be given to advancement, refinement, and application of current and emerging technologies in research on speech physiology and its resultant acoustic output. Promising techniques include magnetic resonance imaging, x-ray microbeam, electromagnetic transduction, ultrasound, and dynamic palatography. These activities will vastly improve the ability to visualize the hidden anatomy of speech as well as provide the foundation for future basic and clinical research.

Microcomputer Applications

The power, economy, and increasing availability of microcomputers hold great promise for an expansion of research and delivery of clinical service. Specially designed software and peripheral hardware allow these instruments to serve as cost-effective, flexible, sophisticated tools for the study of communication disorders and for the clinical assessment and management of many of them. The rapid development and

refinement of software and hardware is making microcomputer technology simultaneously available to research programs and to clinics. As a consequence, progress in research can be readily transferred to clinical application, and clinical experience can be used to modify research objectives. The insightful use of microcomputers can extend a clinician's effectiveness and productivity. In addition, microcomputers can aid greatly in the development of quantitative procedures for the study of speech. Data analyses that required substantial investments of time only one or two decades ago can now be accomplished swiftly with microcomputers. Several software programs are now available for the acoustic analysis of speech, and systems for aerodynamic analysis are being introduced. One microcomputer in a clinical environment can be the means to collect and store data on speech acoustics, speech aerodynamics, phonologic patterns, and language functions. The evaluation of these various systems, however, has not kept pace with their introduction in the laboratory and clinic. For this technology to be used most effectively for both research and clinical purposes, careful evaluations should be made of the accuracy, precision, ease of use, and suitability in various clinical populations of specific systems as they develop.

- There needs to be information maintenance on the substantial numbers of microcomputers now in use in research and service delivery in the field of speech. Standard evaluation protocols should be systematically applied regarding their usefulness in focused applications.

Augmentative Communication Devices

One of the most substantial changes of the decade has been the development of augmentative communication devices and clinical centers devoted to prescribing them and to training in their use. However, while the devices obviously fill a need in the motorically nonvocal, human engineering considerations have obviously been somewhat neglected during this period of rapid growth.

At present, there is no clear picture of the different populations who now use such devices, of their different motor and cognitive limitations, and of their access to adequate training or probable duration of use. Such information is essential for improving the performance of device designs.

Furthermore, detailed information on how clinicians choose devices is needed, what opportunities they have or want for customization, how they evaluate the performance of a device, and how they are informed about client satisfaction.

Use-monitoring software should be developed, so that clinicians will be able to assess their prescriptive successes and failures.

With respect to future developments, formal assessment procedures are needed that will allow the verification of the superiority of a newly developed device to existing systems. Some particular needs appear to be novel sensors of reduced motor control, techniques for processing user input that disambiguate "noisy" motor input (such as "filters" for tremor), and devices that incorporate communication

with environmental control, such as wheelchair steering.

- "Real life" studies are needed of the characteristics of the client population for augmentative communication, their interactions with existing devices, and their unmet needs, for the development of genuinely improved designs.

SUMMARY OF RESEARCH RECOMMENDATIONS

Major Scientific Research Opportunities

Normal Anatomy and Physiology

- Conduct population-based studies of gross structure of the vocal tract.
- Characterize tissue properties of muscles of articulation.
- Study peripheral and central neural mechanisms of the speech system.

Normal Speech Development and Aging

- Conduct large-scale retrospective and prospective studies of speech development in normal children, comparing normal populations with those at risk for communicative disorders.
- Study the effects of normal aging on speech development, so that abnormal signs can be readily detected at any time in the life cycle.

Devices for Speech Analysis

- Develop and apply new techniques for static and dynamic imaging of the vocal tract, including magnetic resonance imaging, x-ray microbeam, magnetometer systems, ultrasound, and dynamic palatography.
- Develop articulatory synthesis techniques.

Major Clinical Research Opportunities

Speech of the Hearing Impaired

- Conduct large-scale studies of the effects of hearing loss at different stages of the life cycle on speech production.
- Investigate factors with major effects on the intelligibility of the speech of the hearing impaired as a function of educational history and residual hearing.
- Develop speech training aids, with protocols for their use. Such aids must deliver articulatory information in a focused manner and assess learning independently for different components of the articulatory system.

Speech Production in Individuals Who Stutter

- Conduct studies focused on discoordination among respiratory, laryngeal, and articulatory systems.
- Investigate the effects of standard therapeutic regimens on various aspects of articulation.
- Investigate stuttering symptom constellations, with an objective of delineating subgroups of individuals.

Speech in Individuals with Craniofacial Anomalies

- Study the clinical history of patients with orofacial anomalies, with careful grouping by genetic syndrome. The full range of communicative defects should be assessed before and after treatment.

Speech in Neurological Disorders of Communication

- Develop standardized assessment protocols, based on perceptual, acoustic, and physiological data, for neurological disorders of communication.

Research Needs Across Populations

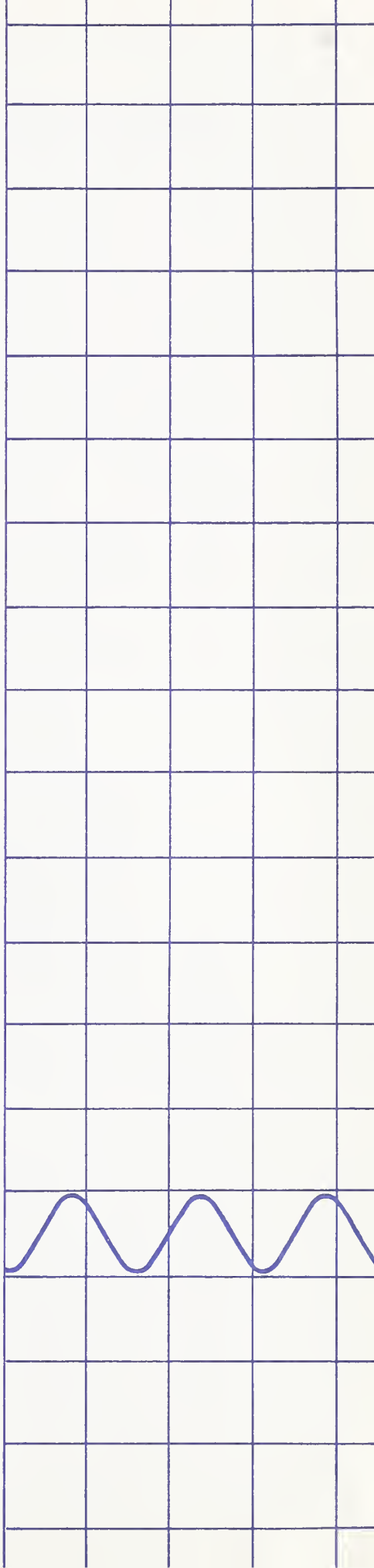
- Develop basic epidemiological information on the prevalence and risk of speech disorders in multicultural populations, with emphasis on nondiscriminatory assessment and effective service delivery.
- Conduct long-term studies of school-age populations assessing relations among speech and language disorders.

Technological Needs Across Populations

- Develop a data base of microcomputers in use in research and service delivery.
- Conduct "real life" studies of characteristics of client populations for augmentative communication, their interaction with existing devices, and unmet needs. Results of such studies should guide device development.

5.

Language and Language Impairments



CONTENTS

OVERVIEW 159

RECENT ACCOMPLISHMENTS 160

Language Development and Disorders in Children 160

Deafness and Language Development 161
Language Disorders in Children 161

Children with Specific Language Impairment 161
Children with Acquired Language Impairment 163
Children with Autism 163

Language Disorders in Adults 164
Speech and Sign Perception 165

Developmental Speech and Sign Perception 165
Speech and Sign Perception in Adults 167

RESEARCH PRIORITIES 168

Language Disorders in Children 168

Relationship of Speech and Language 168
Language Development in Deaf Children 168
Studies of Literacy in Deaf and Language Impaired Children 169
Typologies 170
Processes and Mechanisms 170
Cross-Linguistic and Cross-Cultural Effects 170
Psychosocial Disorders 171
Brain-Behavior Relationships 171
Intervention Strategies 173

Language Disorders in Adults 173

Brain-Language Relations 173
Comparative Language Studies 174
Analysis of Processes Underlying Language Disorders 175
Recovery Processes and Intervention Strategies 176

Speech and Sign Perception 177

In Infants and Young Children 177
In Adults 178
In Normal Subjects 178
In the Hearing Impaired and in Patients with Communication Disorders 180

SUMMARY OF RESEARCH RECOMMENDATIONS 182

Major Scientific Research Opportunities: Children 182

Normal Processes and Mechanisms 182
Deaf and Language Impaired Children 182
Brain-Behavior Relationships 182

Major Scientific Research Opportunities: Adults 182

Brain-Language Relations 182
Behavioral Investigations of Language 182

Major Clinical Research Opportunities: Children 183

Understanding the Nature of Disorders and Atypical Language 183
Intervention Strategies 183

Major Clinical Research Opportunities: Adults 183

Classification 183
Intervention Strategies and Recovery 184

Major Scientific Research Opportunities: Speech and Sign Perception 184

Speech and Sign Perception in Infants and Children 184
Speech and Sign Perception in Adults 184

Major Clinical Research Opportunities: Speech and Sign Perception 185

Assessment 185

5. Language and Language Impairments

OVERVIEW

Language impairments affect children as well as adults and impede economic self-sufficiency, academic performance, and employment opportunities. It is estimated that between 6 and 8 million individuals in the United States have some form of language impairment. In addition to loss of livelihood, these disorders impose social isolation and personal suffering on the affected individuals and place an enormous emotional and economic burden on their families and on society as a whole.

Language impairment is a disorder of comprehension or production sufficient to impair interpersonal communication. These disorders include impairments in the acquisition of comprehension or production of spoken language and may also lead to impairment in reading and writing. In adults and in older children, the impaired include individuals who have lost their previous levels of language competency as a result of brain injury.

In the United States, there are as many as 2 million persons who lost their hearing before they acquired language (the perlingually deaf). A large proportion of these individuals use a form of sign language as their primary mode of communication. A somewhat smaller group uses spoken English exclusively. Many use both. Most perlingually deaf individuals have the potential for normal interpersonal communication and economic self-sufficiency, and those who acquire sign language early have available a full language vehicle for their early cognitive and social development. Nevertheless, they have difficulty acquiring spoken language and in learning to read and write.

The broad goals of research on language disorders are to understand the nature of the various disorders affecting normal language function and to address their underlying bases and the mechanisms and processes that are impaired. The ultimate goal is to develop appropriate intervention and rehabilitation strategies to improve and enhance the communication process for individuals with language disorders. In reaching that goal, it is necessary to understand not only language disorders but also normal language. The understanding of normal language, whether by speech, sign, or writing, provides a basis for comparison in investigations of language disorders. It is critical to understand how language is produced and understood, what its biological and neurological substrates and organizing principles are, how it is learned, and how it is processed by adults.

Disorders of language affect children and adults differently. For the child who from birth does not use language normally or who acquires the impairment in childhood, the disorder occurs in the context of a language system that is not fully developed or acquired. In contrast, for the adult who acquires a language or speech impairment, the disorder affects a fully functional language system. Neurological, physiological, and metabolic differences between children and adults provide particular problems and challenges to the study of language disorders in these populations. As a result, while the broad goals of research on language disorders are similar for children and adults, the research agenda for these two groups is considered separately.

The Task Force addressed three broad issues: language disorders and development in children, language disorders in adults, and speech and sign perception in children and adults. In each of these three areas, recent progress is described, and the highest research priorities for the next three years and beyond are outlined.

RECENT ACCOMPLISHMENTS

Language Development and Disorders in Children

Language learning takes place naturally for most children, in those who have normal hearing and those who are deaf with deaf parents. Recent research has shown that deaf children of deaf parents are not language disordered at all. They use a natural sign language, American Sign Language (ASL), which shares an underlying organization with spoken language, and recent electrophysiological findings show that, in spite of the very different input/output system employed, the same areas in the left hemisphere are involved in language tasks in native signers and speakers of English. Despite the ease with which they acquire this visual communication system, deaf children have inordinate difficulty acquiring reading and writing. Similarly, other children with hearing disorders also have difficulty acquiring reading and writing. In fact, the average high school graduate who is deaf reads at a 3rd to 4th grade level. By studying normal language development, one can gain insights into the normal processes and gather information for comparison with children who exhibit language disorders.

Children with language disorders are a heterogeneous group. Language disorders may be evident in the comprehension and/or production of grammar, vocabulary, and phonology, as well as in the use of language for social discourse. Some language disordered children show a constellation of accompanying behaviors that lead to the clinical label of autism. Many other children exhibit language deficits in spite of normal nonverbal intelligence, hearing, and motor development. Some of these children's language disorders can be attributed to demonstrable brain injury sustained before or after the initial stages of language acquisition. For a larger number of children, the cause of the disorder is not clear.

Until recently, the model for studying children with language disorders was medically oriented. In that model, language disorders in children were related to mental retardation, hearing loss, emotional or behavioral disorders, and neurological impairment. However, children with language disorders fall into many heterogenous groups. It has become clear that language disordered children most often can be assigned to more than one group. Even within a group, their language characteristics are heterogeneous. Children with language disorders may also have concomitant problems (for example, perceptual and motor processing problems) that interfere with language learning. Therefore, it is important that a research agenda address needs in all these groups.

Deafness and Language Development

The terms *deaf* and *deafness* in their most generic sense are applied to children whose hearing impairment is so severe that they are not able fully to acquire spoken language through unaugmented auditory input. In general, this category includes all hearing impaired children who are in special education populations.

Evidence has accumulated indicating that sign language is acquired naturally and with relative ease by deaf children of deaf parents. These findings are especially profound because they indicate that linguistic structural regularities can be processed readily by the human mind, even when they are in a different modality (that is, a three-dimensionally organized phonology and the pervasive use of spatial organization).

Studies of the acquisition of ASL suggest that signers acquiring the language at a later age demonstrate grammatical deviations from more standard ASL, which persist even four or five decades after acquisition has taken place. This finding indicates the existence of critical periods for acquiring sign language that parallel those documented for spoken language.

Studies of deaf children of hearing parents indicate that some of these children may develop rudimentary communication in a gestural mode without any sign language input. Their acquisition of speech tends to be superior to that of deaf children of deaf parents. However, deaf children of hearing parents often display substantial delays in language acquisition and in learning to read.

The dominant pedagogical methodology employed currently in the United States involves the exposure to artificial signing systems, which are intended to represent and model English and to promote the natural acquisition of English grammar. Recent research has shown that deaf children of hearing parents, when exposed only to artificial signing systems, develop idiosyncratic grammatical patterns that reflect neither the structure of English nor of ASL. Their errors are similar to those made by foreigners learning English.

Language Disorders in Children

Children with Specific Language Impairment

Early studies of the neurological status of specifically language impaired (SLI)

children found evidence for frank neurological impairment in only a minority, although limitations in technology might have left the percentage of documented cases artificially low. Neuropsychological studies, however, demonstrated that SLI children performed especially poorly on perceptual and memory tasks, specifically those involving perception of rapid acoustic changes.

Earlier linguistic investigations of SLI children focused on the features of syntax, morphology, phonology, and semantic relations. These studies revealed significant limitations in each of these areas. Although each feature is acquired in a manner approximating normal development (albeit more slowly), the profiles across features often do not match those of younger, normally developing children in that certain features (for instance, morphology) may show an especially serious deficit. Because studies have focused exclusively on SLI children acquiring English, it is not known whether the observed profiles reflect great difficulties with particular grammatical functions or whether they are influenced by the manner in which these properties are marked in English. Several retrospective, followup studies of SLI children suggested that residual problems with language and language-related learning problems may be seen through adolescence and into adulthood.

Research in recent years has begun to address the problem of inadequate subject description. This work has provided a protocol for selecting a prototypic group of SLI children. Furthermore, projects devoted to distinguishing among subgroups of SLI children are now under way.

New technologies (for instance, magnetic resonance imaging [MRI]) are beginning to be applied to SLI children to determine whether previously unidentified neurological disorders might be found in language impaired children. Genetic studies of SLI children have also been undertaken. The preliminary findings from these studies suggest that SLI children are more likely than normally developing children to have other members of the family with present or resolved language difficulties.

Research has continued on the language features giving SLI children difficulty and also on the perceptual and motor abilities of these children. In addition, the lexical (vocabulary) and pragmatic (communicative-conversational) abilities of SLI children have received investigative attention. At this point, sufficient data have been obtained to permit integrating these bodies of evidence.

Prospective longitudinal research on SLI children has begun. The results of these studies indicate long-term deficits in these children. Evidence is accumulating that young SLI children are clearly at risk for later deficits in reading and that SLI and reading disabled children represent overlapping populations.

Several large projects on language intervention are in progress. These have focused on language features such as syntax, the lexicon, and phonology. Studies include comparisons of approaches to treatment.

Children with Acquired Language Impairment

Significant advances have been made in the study of children with acquired brain injury. For some time, it was believed that children sustaining left hemisphere lesions before 1 year of age were much more likely to show higher language ability at a later age than children sustaining lesions after 1 year of age. This does not appear to be the case, at least for language comprehension ability. Although both groups of children subsequently acquire (or reacquire) language, they typically continue to fall behind neurologically normal age-mates on language tasks. Interestingly, these children's difficulties seem to be related as much to limitations in memory as to problems with specific linguistic structures.

Children sustaining right hemisphere lesions also exhibit limitations in language comprehension relative to normal peers. The reasons for this are not yet clear, but preliminary evidence suggests that attentional limitations and visual spatial difficulties hamper these children's performance on language tasks.

Investigation into the relationship between lesion location and language ability has begun. On the basis of the few children with left hemisphere lesions examined to date, it seems that retrorolandic lesions are associated with greater language comprehension difficulties than are prerolandic lesions.

Children with Autism

Recent studies of autistic children have included attempts to identify subgroups within the population, based on different etiologies as well as on behavioral profiles. Furthermore, the contribution of specific behaviors to the degree of severity of autism has been examined. Although communication deficits remain a hallmark of autism, it appears that certain nonlinguistic deficits make a separate contribution to the impaired interpersonal skills of these children. For example, autistic children's poor social skills seem to be more closely associated with their deficient perspective-taking ability in nonlinguistic domains than with their limited ability to comprehend language.

Consistent with the growing view that autism is primarily a disorder of neurological origin, recent research has focused on a number of potential sources of neurological dysfunction. Abnormal averaged brainstem-evoked responses and reduced responsivity to vestibular stimulation suggest that these children may suffer from dysfunction of the brain stem. There is also evidence indicating that, in response to linguistic stimuli, autistic children often show right (rather than left) hemisphere dominant patterns of hemispheric asymmetry. Evidence has also been found by postmortem study and MRI for a previously unsuspected structural anomaly in the cerebellum of many persons with autism.

Studies of the language of autistic children have focused on comprehension strategies, on spontaneous use of syntax relative to comparison groups, and on the utility of augmentative and alternative communication techniques of various types. Clinical trials of drugs that may ameliorate some symptoms of autism are in progress, and a number of promising new pharmacological

interventions have been proposed. Studies of cognitive differences in autistic children have revealed marked differences relative to controls, despite equivalent IQ scores.

Language Disorders in Adults

Disorders of language in adults include total or partial impairment of the capacity to produce spoken language or to understand it; impairment of the ability to read or write; and in the case of the profoundly deaf, loss of the ability to use sign language. The affected clinical populations span a range of etiologies. They include adults who have been rendered aphasic by stroke, brain tumor, infection, or head injury; those who have had developmental language problems of long standing; those who have failed to acquire spoken and written language because of deafness; and those with lifelong autism. In addition, they include individuals in the aging deaf or hearing populations whose loss of language is either an isolated symptom or a component of Alzheimer's disease or other dementias.

Research over the last 10 years has been particularly productive for a number of aspects of brain-related language disorders. For example, with respect to direct correlations between brain and language functions, the road has been cleared for intensive exploitation of refined brain imaging techniques. Already, fine-grained analysis of computerized tomography (CT) and MRI scans has permitted the differentiation of previously unrecognized subtypes of aphasia, the identification of lesions associated with specific deficits, and predictors of recovery or of lasting impairment. Imaging of brain metabolism by positron emission tomography (PET) is leading to the identification of structures involved in such complex operations as naming and reading.

The application of techniques of modern cognitive science has brought about major refinements in understanding the processes underlying language functions that directly affect diagnosis and approaches for remediation. Common mechanisms underlying the sign language of the deaf and natural spoken language have been dramatized by parallels in the effect of brain injury on these two modes of communication. The feasibility of artificial intelligence computer models of language impairment has been demonstrated as a promising technique for testing and describing hypotheses concerning the bases of particular language deficits. Cross-language studies have revealed that apparent symptoms of language disorders are affected by the structure of the language of the speaker.

High points in the remediation of language disorders include the demonstration that profoundly aphasic patients can learn a computerized technique for exchanging information by manipulating visual symbols. Results of pharmacological treatment have shown promise in the amelioration of some of the most severe components of aphasia. Efficacy of specific therapeutic techniques has been demonstrated—among them, the elimination of perseverative intrusions of earlier utterances and the development of adaptive strategies to assist the language impaired to circumvent their limitations.

By building on prior research accomplishments, new studies have focused on imaging (anatomical, morphological, and metabolic) of both normal and brain-damaged subjects. The results are providing a deeper understanding of language-brain relations. Advances in the investigation of the nature of language deficits have continued as a result of developments in linguistic theory as well as in the application to language disordered populations of methodologies and theories derived from cognitive psychology. Further research has explored the effects of brain injury on the processing of language and other cognitive skills in deaf individuals. Remediation techniques continue to make use of state-of-the-art computer applications, and pharmacological treatment continues to be an area of great promise.

Speech and Sign Perception

Human language is accomplished typically through speech and sign. (This area of research is fully discussed in chapter 5.) To understand the nature of speech and sign, it is necessary to know how the systems work when they are fully realized in the adult, and how they come to be fully realized in the developing child. The study of speech and sign perception in children as well as in adults provides converging information about the biology of language, the interaction of biology and environment in the acquisition of this complex symbol system, the extent to which speech is perceived in a way that is similar to other complex stimuli (for instance, nonspeech auditory stimuli and visual stimuli such as objects), and how speech and sign are ultimately organized in the brain. This knowledge forms the essential foundation for an understanding of disorders of speech, sign, and language and for developing appropriate remediation techniques and strategies.

Just as speech is the basic building block of language for the hearing, so sign is the basic building block of language for the deaf. Thus, the study of sign perception is as critical to the study of language in the deaf as the study of speech perception is to the study of language in the hearing. Such study will provide not only insight into the nature of language processing in the deaf but also the foundation for understanding disorders of speech and language that may occur in the deaf. Of critical importance, comparisons of the processes of speech perception and sign perception in normal and language-disordered hearing and deaf individuals provide a unique means of determining those aspects of language that are independent of the mode (signed or spoken) of communication. Such findings may be used to develop appropriate rehabilitation strategies, depending upon the nature of the language deficit.

Developmental Speech and Sign Perception

During the last decade, the understanding of normal infants' processing of speech advanced greatly. New techniques have allowed clinicians to test perception in infants as young as 2 to 3 days of age. The results of studies show that normal infants' abilities to hear differences in speech, like those between individual phonetic units such as the /b/ in "bat" and the /p/ in

"pat," is very good at birth. In the first 6 months, discrimination is present even for sound units that are not present in the infant's own language. Moreover, at least as early as 6 months of age, infants are capable of recognizing a particular speech unit when it is spoken by different talkers using different tones of voice, thus demonstrating an ability that computers have thus far not demonstrated.

The most recent findings in this area show that infants' processing of speech goes beyond that involving a single modality. By 20 weeks of age, and perhaps earlier, infants show a primitive ability to lipread—they detect a match between the auditory presentation of a sound and the sight of a person producing that sound. Normal adults use lipreading to help perceive speech in noisy situations, and research shows that they are strongly influenced, even under good listening conditions, by the visual information obtained by watching a talker's mouth movements. The fact that infants demonstrate simple lipreading abilities at such a young age suggests that they are learning to relate audible and visible speech by watching and listening to adults speak. Hearing impaired or blind infants are less able to learn from this natural experience.

These results of cross-modal perception suggest that infants are very good at substituting information from one sensory modality for another. Deaf infants learning sign language also rely on cross-modal perception, movement of the hands and arms, and processing by the eye. New technology for three-dimensional motion analysis has been developed for the study of sign perception, and investigations are now under way that will allow one to sort constraints imposed by the transmission modality from more centrally determined factors in the perception of the basic building blocks of language.

Recent findings also show that as early as 12 weeks of age, normal hearing infants imitate the speech sounds that they hear. Imitation is the normal way that infants learn to produce the phonetic and prosodic (pitch, intonation, duration) aspects of their native language. The fact that normal infants begin so early to mimic the sounds they hear means that they are already mapping out the relationship between speech movements and sound. Infants eventually learn which speech movements produce sounds that match those produced by adult speakers of the language. Hearing impaired or articulation-deficient infants will not have the same learning experience and, therefore, will not benefit from it to the same degree. Animal models of song learning in birds illustrate the importance of auditory perception to early vocal learning, and suggest that there are "critical periods" for learning. There is little information about such critical periods for speech perception learning in human infants or sign recognition learning in deaf infants, particularly related to the cross-modal aspects of such learning, and the investigation of such critical periods is an important subject for future research.

In another recent finding, there is evidence of a developmental change in the ability to discriminate speech sounds, which reflects an influence of exposure to the infant's native language as early as 12 months

of age. At this time, infants begin to lose the ability to discriminate phonetic contrasts that are not important in distinguishing words in their own native language. Thus, as early as 12 months, infants begin to show a shift from the ability to discriminate the sounds from all languages to a more focused ability to discriminate the sounds from their own language.

Finally, there is new evidence of the need for a good listening environment early in life. This need is illustrated by two findings. First, there are some data to suggest that chronic otitis media in the developing infant, which produces a mild and intermittent hearing loss, may be predictive of later language impairment. Second, there is evidence that normal infants and children need a more favorable signal relative to the background noise to perform at the same level as the adult in speech perception tasks.

Speech and Sign Perception in Adults

Basic to the understanding of disorders of speech and sign perception in adults and speech and sign perception in the hearing impaired is an understanding of normal processes. Research advances in the past 10 years have focused primarily on speech perception in normal persons, and while many advances have been made, scientists are only beginning to understand fully the nature of the speech and sign perception process.

Cross-language studies have shown that, despite the many differences in the sound systems of languages, a basic and small set of clusters of attributes called *features* is used across languages. Sign language perception also seems based on a small set of basic features. Further research has shown that constraints of the auditory system and the physiology of the sound production system help structure the sound inventories of languages, and help define the finite set of features. Although this research is still ongoing, it has already been shown that the acoustic cues used by the listener in perceiving these phonetic features or dimensions vary considerably, with many cues corresponding to a single phonetic unit. Importantly, it has been shown that these cues vary with context and are subject to many sources of variability. Because listeners still perceive a stable phonetic unit despite these many sources of variability, it has been suggested that speech is "special," requiring specialized neural mechanisms.

More recent research findings have indicated that while the acoustic cues for speech are context-dependent, acoustic properties have been identified that are not context-dependent and that remain stable across many sources of variability. These findings have initiated a renewed emphasis on the analysis of the acoustic structure of speech to determine which acoustic properties are invariant and which are not. More sophisticated instrumentation techniques, particularly computer-implemented speech analysis developed in the past decade, have provided a more precise delineation of the acoustic characteristics of the speech signal. In line with the renewed emphasis on the acoustic signal, recent investigations have begun to

explore how the acoustic characteristics of the signal are initially coded neurally as it ascends the auditory pathway. The findings will be very important for the design of cochlear prostheses.

For most people, speech or sign is the way to express thoughts and ideas and to communicate with each other. How speech or sign perception interacts with syntactic and semantic knowledge in language comprehension is not understood to date. Such research is beginning, however, largely through the efforts and cooperation of many different kinds of scientists (speech and hearing scientists, engineers, linguists, and psychologists) contributing a multi- and interdisciplinary perspective to this complex but critical problem. Research findings to date suggest that speech perception affects how persons recognize and access words and also provides critical cues to the syntactic structure of sentences.

Research accomplishments in the past year have built programmatically on the research advances of the past decade. Increased availability of precise instrumentation techniques and use of more sophisticated research methodologies promise to provide a richer understanding of the nature of speech perception and sign perception. The basic properties of the speech signal continue to be elaborated. The nature of the speech perception process and the role of contextual factors in that process is an area of ongoing exploration. The acoustic and phonetic attributes characterizing different languages and how these attributes relate to the sound structure of language continue to be investigated. The auditory basis of speech processing is an ongoing priority.

RESEARCH PRIORITIES

Language Disorders in Children

Relationship of Speech and Language

The relationship of the speech perception and speech production mechanism to language should be investigated. This subject may be approached most readily by studies of normal infants and young children. Specific questions are:

- Are speech perception and speech motor abilities progressively integrated with one another at different developmental levels before language is acquired?
- Is this integration of skills essential to the beginning of language acquisition?

Language Development in Deaf Children

It has been demonstrated that deaf children of deaf parents tend to outperform all other deaf children on a variety of academic and psychosocial measures. A full explanation of this phenomenon is lacking,

although it has been suggested that it stems from the fact that such children have access to a rich and natural communication environment. The patterns of language acquisition in such children are still little understood. Thus, studies are needed of ordinary patterns of acquisition of ASL and its relation to cognitive and psychosocial development among prelingually deaf children of deaf parents. In addition, to understand normal patterns of such development, it is necessary to pursue parallel studies of disordered courses of development of sign language in the same group.

Because deaf children of deaf parents constitute only a small proportion of the population of deaf children (perhaps 5 percent of the total), it is imperative that a better understanding of typical patterns of language development of deaf children of hearing parents be pursued. This knowledge is crucially needed because this group poses the greatest challenge to educational programs. Studies of this group should include:

- Identification and description of patterns of language acquisition of ASL (and related cognitive and psychosocial developmental considerations) when access to a first natural language is delayed or incomplete.
- Identification and description of patterns of language acquisition of children primarily exposed to systems or styles of signing in which the intention is to model or support English speech (for example, signing exact English).
- Identification and description of patterns of language acquisition of children exposed primarily to oral language.
- Studies of the nature and value of language models provided by hearing adults in contact with deaf children. This research should include studies of the nature of productive and receptive aspects of English-like signing.
- Identification of deaf children who do have language disorders and determination if there are differences in perceptual and motor capabilities between normal and language impaired native signers.
- Examination of perceptual and motor processing in deaf children who have acquired spoken language as compared with those who are primarily signers in ASL.
- Studies of approaches and techniques for the training of hearing parents of deaf children in the use of natural sign language.

Studies of Literacy in Deaf and Language Impaired Children

Because problems associated with the acquisition of reading and writing skills are the most pervasive academic and vocational consequences of early-onset language disorders and deafness, and because children with early language intervention and early access to ASL tend to achieve at a higher level than other language disordered or deaf children, studies of the acquisition of English literacy must be included as a primary research objective. This effort should include:

- Studies of the nature of processing of text by successful deaf readers and the

development of effective visual (nonauditory) strategies for the teaching of literacy to deaf and language impaired children.

- Development and testing of new strategies and techniques for teaching English as a second language to deaf children that make use of their capabilities for visual language processing.
- Studies of the impact of closed captioning, telecommunication devices, personal computers, and other technological influences on deaf and language impaired children's literacy skills.
- Studies of the effects of speech processing and literacy in language impaired children and in deaf children who use spoken language primarily.
- Development and testing of appropriate and psychometrically sound tests of psychosocial, intellectual, academic, and linguistic development for deaf children.

Typologies

It is important to determine if language disordered children may be categorized as, for example, verbal agnosic, verbal dyspraxic, autistic, or specifically language impaired. Specific goals are to:

- Develop a rational typology for language impairments in children.
- Determine the extent to which different perceptual and motor disorders are characteristic of subgroups so defined.
- Determine whether or not language impaired children shift from one typology to another as they mature and as their processing strategies change.
- Develop psychometrically sound procedures for the measurement of language, comprehension, and production, to be used in identifying language impairments in children and in classifying these impairments.

Processes and Mechanisms

It is essential to investigate in more detail the underlying processes (sensory, perceptual, motor, cognitive) that subserve language development and disorders. Studies that focus on developing and exploring models that integrate deficits in perceptual, motor, and/or cognitive processes with specific deficits in linguistic processing are needed. Studies of normally developing children are needed for comparative purposes.

- A critical area for further research is investigation of similarities and differences that arise in the processing of sensory information from different sensory systems (auditory, visual, tactile, etc). It is especially important to understand how sensory information from various modalities is used to subserve the development of symbolic representation systems, particularly spoken, signed, and written language.

Cross-Linguistic and Cross-Cultural Effects

Because different languages employ different grammatical structures to convey

the same relationships, it is possible to test hypotheses deriving from theories of language learnability and processing strategies in normal and language impaired children in different language environments. This approach should be pursued vigorously. An especially important question is:

- Do the linguistic profiles of language impaired children vary according to the type of language being acquired, and, if so, do the observed weaknesses reveal a common factor that may be the source of the disorder?

Linguistic differences often accompany cultural differences. The United States has a culturally diverse population. Different cultures may differ in how language impairment is defined. Research in this area seems especially important for the many nondominant cultures within the United States. Children from these cultures can be regarded as multicultural in that they often function within the U.S. society at large as well as in their own culture. One highly relevant question is:

- Can assessment and treatment for multicultural children be improved through a clearer understanding of how the children's language abilities are perceived in the local culture as well as in society at large?

A disproportionate number of children from nondominant cultures are identified as language impaired. The assessment tools used often do not adequately reflect their nonnative language patterns. One critical question is:

- Can assessment and treatment for multicultural children be improved by the development of diagnostic procedures that differentiate disordered language from normal language differences due to a nonstandard dialect of English or to the acquisition of English as a second language?

Psychosocial Disorders

There is still considerable debate about the relationship of language impairment and psychosocial disorders in children.

- Studies should be conducted to determine to what extent language impairment may be secondary to psychosocial disorders and to what extent psychosocial disorders may result from a primary language impairment.

In a special group of children and youths who are first offenders against the law or who have been incarcerated, language disorders and language based-learning disorders appear to be prevalent.

- It is important to determine to what extent failures in communication, academic failure based upon a language disorder, and frustration related to these failures may contribute to the antisocial behaviors of these young people.

Brain-Behavior Relationships

Current research suggests that developmental language disorders, with concomitant perceptual, motor, and memory deficits, are most consistent with a pattern of innate or progressive dysfunction of the dominant temporal association cortex. There may be a functional deficit within the

left hemisphere itself or subcortical projections to this area, an abnormal state of cerebral asymmetry, or disordered interhemispheric integration. The mechanisms by which neurological dysfunction occurs and the precise etiologies that result in developmental language dysfunction are still unresolved. A wide variety of intrinsic and/or extrinsic factors may be responsible for an initial induction of a neurological abnormality: for example, abnormal cell migration, abnormal neurogenesis in regions of the brain where neurons continue to proliferate after birth, or failure of normal subtractive neural processes of neuronal death and axon retraction or disturbance of normal development of myelination of the systems and structures subserving language.

- There is a great need for anatomical, physiological, and metabolic studies of the normally developing brain at different stages of development across the life span. New advances in imaging the brain (anatomical, morphological, and metabolic) are particularly important to apply to studies investigating normal and abnormal development. Particular emphasis should be given to multidisciplinary studies that combine imaging techniques with fine-grained behavioral analysis of processes underlying language development and disorders.

Advances in molecular genetics, coupled with recent evidence of familial transmission of developmental language and learning disability, suggest that further genetic studies would be particularly profitable. Studies should combine state-of-the-art behavioral assessment with molecular procedures. Genetic studies should focus on children with well-defined chromosomal or other genetic abnormalities (for example, inborn errors of metabolism) as well as on children with a well-documented family history of developmental language-learning disorders. Detailed neurological, physiological, metabolic, hormonal, and behavioral testing of family members with a history of developmental communication disorders is also essential.

- The effects of hormones and the immune system on brain development and higher cognitive function is not well established. Recent evidence suggests a possible role of gonadal hormones and the immune system in the development of cerebral asymmetry as well as in functional changes throughout the life span (puberty, menopause). Similarly, recent findings of significant secondary sex-ratio differences (more male than female offspring) in women who were developmentally language-learning impaired also suggests a possible role of these systems in the etiology of the disorder. Studies of the effects of gonadal hormones and immunological dysfunction on brain development and disorders of higher order processes need to be pursued. The development of animal models might be particularly fruitful in this endeavor. Also, studying the effects of hormonal change in language-learning impaired individuals might yield particularly important results.

Intervention Strategies

New intervention strategies, based upon current theoretical models, need to be developed for language impaired children. The strategies should be capable of testing and refining those models. The strategies should also be tailored to meet the needs of individual children; that is, they should address documented, underlying perceptual and motor deficits as well as linguistic inadequacies.

- Efficacy and effectiveness of different intervention strategies should be tested in single-subject studies and in studies of children who clearly represent different typologies of language impairment.
- Animal models should be consulted in developing new intervention strategies for use with severely language impaired children.
- The use of computers and use of recent technological advances (for example, in speech recognition and speech production) should be investigated in intervention with language impaired and deaf children learning oral language.
- The value of assistive devices such as cochlear implants, tactile aids, and visual aids should be assessed in intervention programs using spoken language with deaf children.

Language Disorders in Adults

Research needs for adult language disorders fall into four broad areas:

- A fuller understanding is needed of how language in all of its many aspects is mediated by anatomical structures of the brain and their physiology. The understanding of normal language processes is inseparable from that of the breakdown of language as a result of disruption of structure and function in the brain.
- Comparative studies among various forms of language impairment are needed to reveal common mechanisms. The results should lead to more rational approaches to diagnosis and treatment.
- Functional analysis of processes underlying specific deficits and abnormalities of language use is needed to remedy shortcomings in current classification systems, to sharpen assessment procedures, and to provide potentially more effective rehabilitation techniques. Here, too, an understanding of normal language goes hand in hand with an account of abnormal function.
- Development of new therapeutic tools and outcome studies to evaluate their effectiveness and to track recovery over the long term are needed.

Specific research objectives are discussed below.

Brain-Language Relations

- Individuals vary widely in their recovery of language capacities after apparently similar brain lesions. The mechanism by which recovery takes place and the basis for individual

differences can now be investigated more effectively by PET scan imaging of brain metabolism. The role of the right side of the brain and of residual function of structures in the left brain needs to be understood.

- While detailed correlation between the breakdown of motor speech planning and structural brain lesions has been demonstrated, other potentially localized language skills, such as speech sound discrimination and oral word comprehension, are still inadequately mapped. The line between processes that are closely related to well-localized structures and those that are broadly distributed in the language zone should be further defined by clinico-anatomic study. Such an understanding will help direct therapeutic efforts to functions that have the best prospects for recovery.
- Cases presenting well-defined forms of selective impairments are of vital importance for delimiting structure-function relationships in the brain. Such cases must be identified so that they may be subjected to particularly close analysis of brain abnormalities by imaging and electrophysiological techniques, as well as to careful analysis of functional impairment.
- The role of various neurotransmitter systems in the mediation of particular language operations needs to be further explored. The potential for pharmacological treatment follows directly from such understanding.

Comparative Language Studies

Research on language in the deaf and on language disorders in hearing adults is greatly enhanced by comparing performance between different subject populations. Such comparisons provide additional insights into the nature of language processing in each of these groups. On the one hand, generalizations about the specificity of patterns of language dysfunction as attributable to the particular etiology of the group can be considered, and on the other, differences and similarities between the groups may provide information about the underlying bases and mechanisms responsible for the various language disorders. The following comparative studies are of particular interest and are likely to provide critical insights:

- *Studies of the normal elderly.* For spoken, written, and signed language, comparatively little is known about how—and even if—language is modified as individuals normally grow older. Therefore, the study of language in the healthy elderly is of interest in its own right. In addition, there will be little valid progress in understanding disordered language in the elderly without a normative data base against which to compare and contrast. A significant research priority, then, must be the study of language in normal aging.
- *Comparison of right- and left-hemisphere-damaged patients.* While left-hemisphere-damaged patients often recover some or most of their language function, it is not clear whether such recovery reflects left hemisphere mechanisms or the

involvement of right hemisphere mechanisms. The question is: What role does the right hemisphere play in language processing? Comparative analysis of left- and right-brain-damaged hearing subjects will help to determine the extent to which the right hemisphere may be involved in language use. For deaf signers, comparative analysis of the selective impairments that are contingent upon right- and left-brain damage have different, but equally important, theoretical implications. Because signing involves complex linguistic properties conveyed largely through spatial mechanisms, studies of such patients are especially important for issues in determining hemispheric specialization in humans.

- *Specific versus generalized brain damage.* As more explicit and theory-driven descriptions of the disordered language of focally brain-damaged patients (such as those with aphasia) have emerged, it has become clear that differences exist between these language disorders and those that accompany more general brain dysfunction; that is, the language impairments observed in the various progressive dementias and those observed in traumatic brain injury. Clearer descriptions, comparisons, and contrasts among all these language problems can aid in differential diagnosis and provide guidelines for appropriate intervention. Further, understanding the relationships between language disorders and their underlying pathologies can increase the understanding of brain-behavior relationships. For these reasons, comparative studies of language disorders in these conditions should be undertaken.
- *Multilingual-bilingual research.* The population of the United States is becoming increasingly multilingual, and as this population ages, there will be an increasing number of multilingual individuals with language disorders. Research is needed in the bilingual population to further the understanding of deficits and recovery and rehabilitation in aphasia and of degenerative processes in dementia and other conditions that lead to decline of language function. Research in this domain will, like cross-language studies, offer informative tests of hypotheses concerning the representation of language structure and processes in the brain.

Analysis of Processes Underlying Language Disorders

The language impairments that result from brain damage reflect the dysfunction of one or more of the mechanisms that subserve normal language, whether spoken, written, or sign, including perceptual, motor, cognitive, and specifically linguistic mechanisms. For example, the normal ability to read involves visual perceptual mechanisms, mechanisms that control attention to the relevant material on the page, memory components that store knowledge of words, and linguistic mechanisms that compute the meaning of sentences. Damage to any one of these

mechanisms will result in a reading impairment, the specific form of impairment being determined by the particular mechanism that is damaged. Thus, a deep understanding of the nature of the various forms of language disorders consequent to brain damage crucially depends on knowledge of normal language processes and their disruption. There are several important research opportunities in this area.

- Research should be focused on extending recent progress in the cognitive sciences to an understanding of the structure of normal language processing.
- Research should be directed to the improvement of typology of language disorders through the detailed investigation of the linguistic, perceptual, motor, and cognitive mechanisms that are affected by brain damage.
- Research should be directed to distinguishing between language disorders that result from deficits in specific language mechanisms (such as syntactic processing) and disorders that result from deficits in more general cognitive mechanisms (such as attention).
- Research should be directed to distinguishing between (among) forms of dysfunction in oral/aural, written, and sign language. Research that focuses on selective deficits in one language modality and on comparative analysis of deficits in different language modalities should be encouraged.
- Research should be directed to elucidating the relationship between language-specific deficits and perceptual, motor,and cognitive deficits in complex language disorders.
- Comparative analyses of disorders in different languages should be undertaken. Languages differ in ways that can help reveal the linguistic and cognitive mechanisms that subserve language processing. For example, the comparative analysis of reading and writing disorders in speakers of languages with alphabet script (English, Spanish) versus speakers of languages with other script forms—ideographic (Chinese), syllabic (Japanese), or phonetic (Korean)—could provide crucial evidence concerning the organization of the reading and spelling processes.
- An emerging research opportunity is the application of Artificial Intelligence technology for the simulation of language disorders. The development of computer models of normal language processing that can be experimentally lesioned to generate particular forms of language dysfunction should provide an important new tool for evaluating hypotheses about the nature of the linguistic and/or cognitive mechanisms that underlie language disorders. Complex models are particularly important since there is no possibility for the development of suitable animal models to study language disorders. Research in this emerging area should be strongly encouraged.

Recovery Processes and Intervention Strategies

Understanding the relationships among patient characteristics and intervention

techniques is vital to appropriate selection of patients and to refinements of promising techniques.

- Knowledge about spontaneous recovery from aphasia has grown over the past 10 years, and there are somewhat more reliable ways to predict the course of recovery. However, scientists remain unable to predict with reasonable certainty who will recover and the extent to which recovery can be augmented by clinical intervention. In addition, investigators are only beginning to understand the potentially beneficial effects of pharmacological agents upon the recovery process and on recovery augmented by clinical intervention. More research in this area is needed.
- Little is known about predicting the course of progression of disorders where deterioration instead of improvement is the natural course—for example, in Alzheimer's disease. There are few intervention studies of such disorders, either in conjunction with pharmacological agents or without them. This is an area in great need of study.
- The degenerative disorders illustrate a need not only for clinically innovative therapeutic strategies but also for new research methodologies that would permit more valid evaluation of outcome. The development of both innovative and less restrictive new single-subject and group methodologies is encouraged.
- New intervention strategies for working with aphasic and head-injured individuals are needed, particularly strategies that capitalize on the emerging knowledge about language processes or that provide aphasic individuals with workable compensatory mechanisms.
- Research is needed to develop and evaluate alternative and augmentative communication systems for adults with language deficits. Adults severely impaired in understanding and producing language may be capable of effective communication by alternative means, including computer-based systems, when these techniques make demands that are within their preserved abilities.
- There is little doubt that a host of individual factors, ranging from symptoms to age and medical status, affect responsivity to treatment. Social and cultural factors also have potential for influencing treatment. Yet, the way these factors (individually or in combination) affect outcome is not clear and must be accounted for in intervention.

Speech and Sign Perception

In Infants and Young Children

Infants' perception of speech can be assessed early in life, and this ability offers the potential opportunity to see differences between individuals that may be predictive of either an existing hearing impairment or of later speech and/or language disorders.

- It is very important to standardize tests of speech perception in normal infants and young children, so that the degree to which impairments of speech

perception are predictive of language disorders can be assessed.

- The findings on young infants, especially those involving categorization, lipreading, and imitation, need to be extended to see when these abilities begin to show the effects of exposure to a specific language. This effort will require extending the tests on young infants to older age groups, particularly those between 1 and 3 years of age.
- There is a strong need to investigate whether there are critical periods in infant learning, particularly for the learning of the cross-modal connections needed for lipreading (auditory-visual) and for imitation (auditory-articulatory).
- Studies relating sign perception to speech perception are now feasible and are very important to understanding whether language processing is similar, regardless of the modality through which it is acquired.
- There is a need to relate infants' early acquisition of phonology, assessed through tests of speech/sign perception, to the acquisition of the higher levels of language, such as the acquisition of words (lexicon), word meaning (semantics), and grammar (syntax). Multidisciplinary approaches are needed to accomplish this research. Experts in developmental speech perception, developmental sign perception, developmental speech production, general cognitive development, and developmental linguistics are needed to describe the full linguistic competence of the developing child and to determine the extent to which that competence is impaired in language disordered populations. Such an approach is vital to the early diagnosis and treatment of language disorders.

In Adults

Research on adult speech perception has two primary objectives. The first is to articulate more fully and to explicate normal processes for speech perception. The second is to understand on the one hand how and specifically in what ways speech perception processing is affected by hearing loss, and on the other to investigate systematically the extent to which speech perception disorders underlie or contribute to language disorders. Understanding the role that speech and sign play as the vehicles for language communication and for higher levels of language processing is a long-term goal. By comparing speech perception to sign perception, it will be possible to understand better the processes of language perception in general and characterize the bases of language disorders.

In Normal Subjects

- It is essential to understand the nature of the speech signal itself—how it is generated, how it is perceived, and how the processes of speech perception relate to those of speech production. These findings will not only provide an integrated view of speech processing but should also provide a means of exploring the underlying bases of speech disorders.

- It is essential to identify those acoustic cues or properties that are invariant and those that are context-dependent, and to determine how these cues are utilized across languages. Such results should help to determine those cues or properties that may be more vulnerable to neurological insult and those that may be more resistant.
- It is essential to investigate the auditory transformations of complex signals like speech to determine how this processing structures and ultimately shapes human speech. Such results may provide critical information on how neurological damage at different points in the auditory system may affect the processing of speech. There are two ways of exploring this question. One makes use of computer modeling based on psychophysical results in humans, and the second makes use of animal models for mapping out the auditory transformations of the speech signal.
- Competing models for the speech perception process need to be more explicitly detailed so that they can be experimentally evaluated. Such findings allow investigators to converge on the correct account of speech perception mechanisms. Moreover, theories provide a framework for posing research questions, for guiding research programs, and for establishing research strategies and priorities.
- It is essential to investigate both sign perception and sign production. Sign language provides a unique vehicle for analysis of language production, because movements of the hands and arms are directly observable in sign. New technologies should be utilized that allow the three-dimensional analysis of sign articulation. Invariant signed properties should be identified with the aim of delineating factors that shape the form that language takes in the visual modality. These studies should provide an important reference point for the investigation of sign disorders due to neurological damage. Such studies are essential because they can illuminate aspects of the processing mechanisms and underlying neurology of sign.
- Further research needs to focus on speech perception as it relates to lexical access and to sentence and discourse processing. It is critical to consider speech perception beyond extracting the phonetic units of speech. While this effort is long-term, preliminary research is promising, and further important advances could be made within a few years. Critical questions include how speech processing affects or influences word recognition, lexical access, syntactic parsing, disambiguation, and ultimately sentence and discourse understanding.
- A critical area of research is the investigation of speech perception in the elderly. Little is understood about the speech perception abilities of the normally aging individual. Earlier research indicates that the elderly, in the presence of normal hearing, do show changes in their linguistic skills, and

these changes affect their language abilities. It is essential to understand whether changes in speech perception occur with aging, and if they do, to assess the extent to which they affect language comprehension skills. Moreover, to understand language disorders in the elderly, it is necessary to compare their performance with appropriate control subjects. Such subjects would be normally aging individuals.

- It is essential to understand the nature and processes involved in sign perception of the deaf. Such investigations need to include the study of the basic properties of sign, how sign information is integrated with higher levels of language (including lexical, morphological, and syntactic cues), and how sign perception ultimately relates to sentence and discourse understanding. Such research will provide insights into language processing of the deaf. A comparison of the nature of sign perception and speech perception will also provide critical information on the relation of language to the mode by which it is communicated—that is, by spoken language or by sign.

In the Hearing Impaired and in Patients with Communication Disorders

- It is essential to investigate how the perception of the acoustic cues of speech are affected by hearing loss. First, it is important to map out which cues are more vulnerable than others and to determine their effects on the phonetic structure of speech. Second, it is critical to ascertain how speech perception interacts with the perception of lexical and syntactic cues and, ultimately, influences the comprehension of auditory language. Such results will also help in the development of appropriate auditory prostheses.
- It is essential to develop suitable methodologies for exploring speech perception in adult patients with language disorders. While there have been attempts to investigate speech perception in adults with language disorders, progress has been hampered by methodological limitations. In particular, current methodologies require that the patients have unimpaired or minimally impaired auditory language skills. In order to be able to test patients with auditory language comprehension problems, new techniques need to be developed.
- In adult patients with language disorders, it is essential to determine the extent to which impairments in speech perception affect higher levels of language. It is possible that speech perception impairments affect lexical or syntactic processing, and thus contribute to auditory language comprehension deficits. Appropriate intervention strategies can be developed only when the underlying basis of language disorders can be determined.

- It is essential to understand sign perception in deaf individuals who become aphasic or who sustain other cognitive impairments subsequent to brain damage. Such study will provide further insight into the nature of language in the deaf. It will also provide information on the nature of sign as a vehicle of language communication for the deaf, and will help to determine the bases of language disorders in neurologically impaired deaf individuals.
- To understand perception of a sign language, it is necessary to investigate basic and higher level processes underlying vision. Sign language makes structured use of space and movement, and processing sign language requires the processing of complex dynamic arrays. It has been demonstrated that the perception of motion differs in deaf and hearing individuals, and investigations should be directed to understanding motion and form processing in deaf signers and hearing individuals, how such visual-dynamic information is processed linguistically, and how disorders of motion and form processing contribute to sign processing deficits.

* * * * *

The scope and number of problems outlined above are clearly beyond the expertise of a single discipline, and a broad range of professionals, from physicians and psychologists to linguists, audiologists, and speech-language pathologists, are active in research and intervention with language impaired or deaf individuals.

By its very nature, the study of language is a complex enterprise, requiring the expertise of a variety of professionals with diverse training. Language impairments and deafness compound or complicate the problem, so that not only basic research but also clinical training is required by the experts who are expected to conduct the requisite research and develop intervention strategies. It is clear that multidisciplinary approaches to these problems can best be achieved by NIDCD National Research and Training Centers. (See chapter 7.)

There is an alarmingly small number of qualified researchers and professionals in this area, which is reflected by the small number of funded grants (see chapter 7, "Research Training and Research Personnel Resources"). That fact, along with the magnitude of the necessary research outlined here, suggests that such centers would be valuable in stimulating training and research in the area of language and language impairments.

SUMMARY OF RESEARCH RECOMMENDATIONS

Major Scientific Research Opportunities: Children

Normal Processes and Mechanisms

- Determine how sensory information from several modalities is used by normal children in developing symbolic representation in language and literacy.
- Determine to what extent the integration of speech perception and speech production (motor) skills and psychosocial skills is an essential precursor to language development in normal children.

Deaf and Language Impaired Children

- Compare perceptual and motor processing patterns of language acquisition in deaf children acquiring spoken language and the language of signs.
- Develop appropriate *normative* and *criterion-referenced* psychosocial, intellectual, academic, and linguistic development appropriate for use with deaf and language impaired children.

Brain-Behavior Relationships

- Use imaging techniques and genetic studies to elucidate relationships between areas of brain functioning and language-related processes and mechanisms.
- Study the effects of gonadal hormones and the immune system on brain development in relation to language-related processes and mechanisms.

Major Scientific Research Opportunities: Adults

Brain-Language Relations

- Use refined neuroimaging techniques (both anatomical and metabolic) and electrophysiological techniques combined with analysis of functional impairment to determine how language is mediated by anatomical structures of the brain.
- Explore the role of various neurotransmitter systems in the mediation of particular language operations.

Behavioral Investigations of Language

- Determine the mechanisms involved in normal language processing in young adults and in the aging process, using theories and methods of cognitive science.

- Identify the processes underlying language disorders in patients with aphasia or dementia, including perceptual, motor, cognitive, and specifically linguistic mechanisms.
- Conduct comparative studies of different clinical populations to reveal common mechanisms and differences, such as language disorders in hearing and deaf subjects, right- and left-brain-damaged subjects, language disorders subsequent to specific versus generalized brain damage, and language impairments in speakers of different languages.

Major Clinical Research Opportunities: Children

Understanding the Nature of Disorders and Atypical Language

- Identify and describe patterns of spoken language acquisition and acquisition of American Sign Language (ASL) in deaf children in relation to cognitive and psychosocial development, to language models provided (artificial or natural), and to perceptual and motor processes, and also when access to a first natural language is delayed.
- Develop the means for identifying language disorders in deaf children and normal hearing children from different cultures, and develop a rational typology for language impairments in affected children.
- Describe the perceptual/motor characteristics associated with different types of language impairment.
- Disambiguate cause and effect in children with both psychosocial disorders and language or language-based learning disorders.
- Look for common factors as revealed by linguistic profiles of language impaired children from different language-speaking communities.

Intervention Strategies

- Design and evaluate intervention strategies that:
  - are theoretically motivated;
  - are tailored to different subgroups of language or hearing impairment; and
  - include the use of assistive devices and of alternative (intact) modality sensory processing.
- Assess how best to involve parents in intervention with deaf and language impaired children.

Major Clinical Research Opportunities: Adults

Classification

- Improve the typology of language disorders through investigations of the linguistic, perceptual, motor, and

cognitive mechanisms that are affected by brain damage.

Intervention Strategies and Recovery

- Develop intervention strategies for working with aphasic and brain-injured individuals, particularly strategies that capitalize on knowledge about normal language processing or that provide language impaired individuals with compensatory mechanisms, such as augmentative communication systems.
- Investigate effects of pharmacological agents upon the recovery process and on recovery augmented by clinical intervention in aphasia.

Major Scientific Research Opportunities: Speech and Sign Perception

Speech and Sign Perception in Infants and Children

- Investigate the ontogeny of speech perception, focusing on categorization, lipreading, and imitation in relation to critical periods and exposure to language.
- Study the relation of sign perception to speech perception with a focus on understanding whether language processing is similar regardless of the modality through which it is acquired.
- Relate early acquisition of phonology, assessed through tests of speech/sign perception, to the acquisition of higher levels of language such as acquisition of words (lexicon), word meaning (semantics), and grammar (syntax).

Speech and Sign Perception in Adults

- Characterize the basic properties of speech and sign—how speech and sign are generated, how they are perceived, how they vary cross-linguistically, and how the processes of speech and sign perception relate to those of speech and sign production respectively.
- Investigate basic and higher level processes underlying audition and vision, to aid in understanding the perception of speech and the perception of sign.
- Define how speech and sign perception relate to higher levels of language, including words (lexicon), word meaning (semantics), grammar (syntax), and discourse.
- Investigate how the perception of speech is affected by aging, and following hearing loss or brain damage.

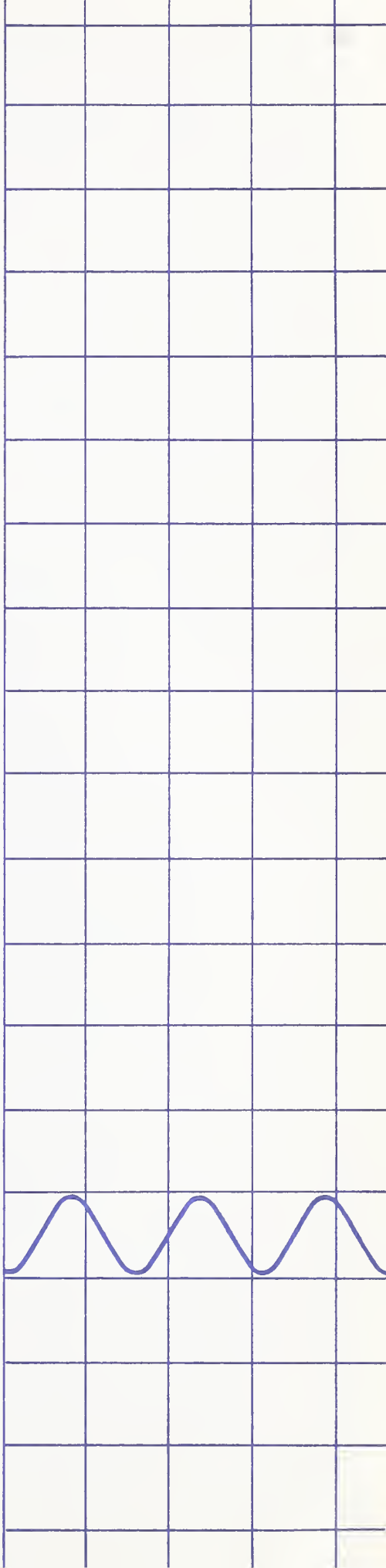
Major Clinical Research Opportunities: Speech and Sign Perception

Assessment

- Standardize tests of speech perception in normal infants and young children to assess the degree to which impairments of speech perception are predictive of language disorders.
- Develop methodologies for exploring speech perception and sign perception in adult patients with language disorders.

6.

Smell, Taste, and Touch



CONTENTS

SMELL AND TASTE 193

Overview 193

Prevalence, Incidence, and Costs 193

Recent Accomplishments 195

Neurobiology of the Chemical Senses 195
Clinical Disorders of the Chemical Senses 197

Program Goals 198

Basic Biological Area 199

Plasticity, Including Development, Regeneration, and Aging 199
Access to the Brain Through the Olfactory Nerve 199
Receptor Mechanisms and Signal Processing Throughout Central Systems . . 200

Clinical Area 200

Epidemiology of Taste and Smell Disorders 200
Development of New Diagnostic Techniques 200
Affective Dimensions of Taste and Smell 200
Associations Between Chemosensory Disorders and Altered Food Intake in Aging and in Chronic Illnesses 201
Effects of Environmental Agents on Chemosensory Function 201
Clinical Trials for Treatments of Chemosensory Disorders and Rehabilitative Strategies 201

Scientific Research Opportunities, Strategies, and Priorities 201

Basic Study of the Chemical Senses 201

Plasticity, Including Development, Regeneration, and Aging 201
Access of Infectious Agents and Toxins to the Brain Through the Olfactory Nerve 204
Receptor Mechanisms and Signal Processing 205

Clinical Study of the Chemical Senses 210

Epidemiology of Taste and Smell Disorders 210
Development of New Diagnostic Tests 212
Affective Dimensions of Taste and Smell 213
Association Between Chemosensory Disorders and Altered Food Intake in Aging and in Various Chronic Illnesses 214
Effects of Environmental Agents on Chemosensory Function 216
Clinical Trials to Evaluate Potential Treatment Protocols and Rehabilitative Strategies 217

Research and Training Centers 218

TOUCH 218

Overview 218
Recent Accomplishments 219
Program Goals 220
Scientific Research Opportunities, Strategies, and Priorities 220

Basic Study of the Tactile Sense 220

Tactile Sensory Coding and Cortical Representation 220
Touch and Its Interaction With Other Senses 221
Development of Tactile Stimulators 221
Haptics 222

Clinical Studies of the Tactile Sense 222

Tactile Sensory Aids 222
Testing of Tactile Sensitivity in the Mouth 223

SUMMARY OF RESEARCH RECOMMENDATIONS: SMELL AND TASTE 223

Major Scientific Research Opportunities 223

Plasticity, Including Development, Regeneration, and Aging 223
Access of Infectious Agents and Toxins to the Brain Through the Olfactory Nerve 224
Receptor Mechanisms and Signal Processing Throughout Central Systems 224

Major Clinical Research Opportunities 224

Epidemiology of Smell and Taste Disorders 224
Development of New Diagnostic Tests 224
Affective Dimensions of Taste and Smell 224

Associations Between Chemosensory Disorders and Altered Food Intake in Aging and in Various Chronic Diseases 225
Effects of Environmental Agents on Chemosensory Function 225
Clinical Trials to Evaluate Potential Treatment Protocols and Rehabilitative Strategies 225

SUMMARY OF RESEARCH RECOMMENDATIONS: TOUCH 225

Major Scientific Research Opportunities 225

Tactile Sensory Coding and Central Representation 225
Touch and Its Interaction With Other Senses 225

Major Clinical Research Opportunities 226

Development of Tactile Stimulators 226
The Haptic Exploration of Objects and Surfaces 226
Tactile Sensory Aids 226
Testing of Tactile Sensitivity in the Mouth 226

6.
Smell, Taste, and Touch

SMELL AND TASTE

Overview

The chemical senses, olfaction and taste, play crucial roles in the detection and identification of chemical stimuli in the environment. These stimuli include foods, environmental chemicals and pollutants, hazardous vapors and gases, as well as pleasurable scents and fragrances. Thus olfactory and taste deficits present serious problems because they can eliminate the ability to recognize and the motivation to consume nutrients; they can eradicate the ability to detect toxins in the home and workplace; and they can decrease the pleasure derived from interpersonal interactions, from meals, and from "just smelling the flowers" in the world around. In addition to the occurrence of chemosensory deficits in themselves, abnormalities in chemosensory responses frequently accompany and even signal the existence of several diseases or unhealthy conditions, including obesity, diabetes, hypertension, malnutrition, and some neurological conditions such as Alzheimer's disease.

Prevalence, Incidence, and Costs

Information about the prevalence and incidence of disorders affecting the chemical senses has proved difficult to assemble. Historically, the best estimates of prevalence, which could not be considered either valid, reliable, or truly representative, were based on extrapolations from limited clinical referral data. In the late 1970s, the opinion of many experts was that more than two million adults in the United States had a chemosensory problem. Often clients expressing concerns about taste and smell were given psychiatric referrals, or told that the problem would go away. So there was not a concerted effort to document or study chemosensory problems.

These earlier rough estimates were significantly improved in 1987 for the olfactory sense following an international population survey conducted by chemosensory scientists with the help of the National Geographic Society. Using a "scratch-and-sniff" format test that was developed with the support of NIH grants, the Society invited readers of its magazine to identify a series of six odorants and to answer several questions about the sense of smell. More than one million people responded. Although this was a nonrandom sample (the readership tends to oversample those of higher socioeconomic status), the results are

nonetheless noteworthy. Highlights of the survey include the following findings:

- Two of every three respondents reported temporary losses in their sense of smell, and those losses often wereassociated with colds, influenza, sinus conditions, and allergic rhinitis.
- Although one-half of the respondents could detect all six test odorants, 1 percent of the respondents could not smell three or more of the odorants. Nearly all could smell banana, but only one-half could identify it.
- More than one-third of the respondents could not detect the sweat-like smell of one of the odorants. Also, nearly one-third could not detect the musk-like smell of another odorant. Thus there may be specific anosmias that have never been characterized.
- Age factors were important. Although the ability to detect smells continues well into the sixth decade, the overall sensitivity to smells begins to decline in the second decade and deteriorates appreciably with age for a significant number of individuals. An unexplained and disturbing finding was that a substantial segment of the elderly show a loss in the ordinary negative reaction to the foul-smelling mercaptan added as a warning agent to natural gas.
- Consistent with earlier findings, smell losses were associated with smoking. Females of all ages identified smells slightly better than did males. The common belief that pregnancy increases sensitivity to odors was not supported by the actual intensity ratings and smell detection tests.

Data of this kind are not currently available for taste. However, with the creation of clinical centers for taste and smell disorders, information has begun to accumulate.

Even the limited, available epidemiologic information is proving highly valuable as biomedical researchers and clinicians try to better understand chemosensory disorders and to assess their impact on the U.S. population. Although extensive systematic data are not available, smell and taste disorders certainly affect diet. While many individuals with olfactory and gustatory deficits somehow seem to maintain nutritionally adequate diets, marked changes in body weight occur in some. In addition, some individuals need to increase their intake of sweets, salt, and spices to derive pleasure from eating, possibly exacerbating chronic health disorders such as obesity and hypertension. Further, losses in the capacity to identify particular odors may likely interfere with an individual's ability to detect toxic substances and reject spoiled or contaminated foodstuffs. How often this loss results in illness has not been assessed.

Another important concern that arises when individuals develop chemosensory losses is the psychological impact for such individuals. Reports are anecdotal, but the severity of impact varies widely, and responses can range from stoicism among some individuals to quite serious complaints, including suicidal impulses, among others.

In addition, it is important to realize that the problems associated with

chemosensory deficits in themselves can be greatly exacerbated when these deficits are combined or associated with chronic illnesses such as cancer, renal problems, or liver disease. In such situations, taste and smell disorders can contribute to problems of anorexia and cachexia, and thereby place individuals at additional risk for nutritional problems.

Although a few methods for evaluating chemosensory sensitivity and losses are available, refinements and improvements are needed. For example, a scratch-and-sniff test now provides an accurate and readily administered suprathreshold odorant identification test. A quantitative threshold test, in which odorants are dispensed from squeeze bottles, may be developed for use in physicians' offices. This would permit the physician to quantify the loss and monitor changes over time.

Refinements in gustatory sensitivity tests are also under development. For instance, refinements that permit measurements of taste sensitivity over different areas of the tongue are proving useful for detecting and localizing not only gustatory deficits but also other forms of neurological damage.

The cost of chemosensory dysfunctions cannot be determined with certainty until better epidemiological data are available, but the impact of these dysfunctions on daily life clearly cost the U.S. population a considerable amount both in dollars and in suffering.

Recent Accomplishments

Neurobiology of the Chemical Senses

With the application of modern neurobiological methods to studies of chemosensory transduction, and increased use of various unicellular organisms and invertebrate species, substantial advances have been made in understanding the events of stimulus access to and interaction with receptors. Investigation of the role of the receptor microenvironment has led to identification of odorant-binding proteins that apparently transport stimulus molecules to the olfactory receptor membrane. Although comparable proteins in the saliva that bathe taste buds have yet to be described, the intimate association between saliva secreted into clefts of papillae on the posterior tongue and the taste buds in these papillae is providing clues about interactions between taste sensation and salivary secretion. Insights about transduction mechanisms themselves have derived from patch clamp studies of olfactory and gustatory receptor membranes. There are multiple lines of evidence now indicating that olfactory transduction depends in part on activity of adenylate cyclases. Moreover, there may be broad, evolutionary-based molecular similarities between the olfactory and visual transduction systems. In both smell and taste, the role of stimulus transport through ion channels in receptor membranes has become increasingly apparent. In the taste

system, there is evidence for involvement of sodium and potassium channels in salt taste transduction mechanisms. In contrast, transduction mechanisms for sugar reception apparently are quite different and involve stimulus binding with specific receptor proteins. Continued application of techniques to study single receptor cell neurophysiology should ensure more rapid advances in understanding chemosensory transduction. Methods to characterize function of single receptor cells and subsequently label the cells for morphological characterization will yield information about specific cell types and their roles in chemosensation.

Central nervous system (CNS) processing of taste and olfactory signals is extremely complex, involving circuits with numerous afferent and efferent systems as well as various areas of the brain. Studies of such central pathways have made apparent the direct links of taste and smell with areas of the brain involved in feeding, drinking, swallowing, breathing, and sexual activity. Data are emerging on the myriad of neurotransmitters in olfactory pathways and on the variety of central neuron morphologies in smell and taste circuits. However, the knowledge about details of these connections and the microcircuitry and neurotransmitters involved in their function will yield only to continued neurophysiological studies and application of immunocytochemistry and single-cell labeling and reconstruction techniques.

The importance of the chemosenses in basic biological, life-sustaining functions is indicated by the early, intrauterine appearance of taste and smell receptors in mammals with a lengthy gestation, including humans. Furthermore, several studies have demonstrated the intrauterine emergence of taste and smell *function*. In animals with relatively long gestations, the taste system is functional at peripheral and central neural levels for at least the last one-half of gestation. However, neural taste responses change during development from prenatal to postnatal stages and long after birth. Altered neural taste responses derive partly from anatomical rearrangements of the taste buds and their innervation during early developmental stages. Not surprisingly, during these periods of neurobiological alterations, the taste system is susceptible to modification by environmental stimuli. For example, diets low in sodium chloride fed to pregnant animals and their newborns alter the development of taste responses to salt. In the olfactory system, intrauterine exposure to chemicals apparently can alter the development of neural pathways involved in processing these odors.

Not only is there early developmental plasticity in the chemosensory systems, but the opportunity for changing function and structure continues throughout life. In the gustatory system, modified epithelial cells that form the taste buds are replaced about every 10 days. This process of cell death and renewal presumably requires remodeling of synapses in the taste buds, to maintain some "order" in neural connections and in sensation. Recent studies indicate that single taste fibers innervate a particular morphological type of taste cell, not heterogeneous cell types. So specific types of connections are sustained even though cell turnover occurs. Olfactory receptor neurons

apparently undergo a cycle of turnover and replacement, of about 30 days in some species. These are the only known neurons with the capacity to replace themselves throughout life. Future understanding of the precise mechanisms that regulate these chemoreceptor cell turnover processes will require concerted efforts at the cellular and molecular levels. This understanding will clarify emerging evidence that auditory hair cells also have some regenerative capacity.

In the gustatory system, the taste buds are apparently maintained through an interactive process with innervating gustatory fibers; the buds degenerate when the peripheral innervation is cut and they regenerate on reinnervation. Such cycles of degeneration and regeneration are associated with recovery from trauma and certain clinical treatments such as chemotherapy and radiotherapy. Not surprisingly, this is coupled with loss and return of chemosensation and with aberrant chemosensation that can lead to altered perception and feeding. Systematic studies of changes in chemosensation in these situations are lacking, however, and an understanding of basic processes is needed to plan better management of compromised patients.

Clinical Disorders of the Chemical Senses

The establishment of the first taste and smell centers has produced improved diagnosis of chemosensory dysfunction, data on significant etiological factors, and preliminary tests of possible therapies.Diagnostic improvements have taken different directions for taste and olfaction. In olfaction, considerable progress has been made on the use of odor identification as a screening test for olfactory losses. One identification test is in the form of a "scratch-and-sniff" booklet. Its form makes it easy to use in a physician's office and to distribute to large groups. Another identification test uses actual household substances presented in small jars. This kind of test is inexpensive and easy for any interested clinician to assemble. Identification tests are used to classify patients into three groups: anosmic (virtually no ability to smell), hyposmic (reduced smell function), and normosmic (normal).

Ageusics (complete taste losses) are rare, so taste tests are focused on distinguishing the hypogeusics (reduced taste function) from those with normal taste. Reduced taste function can be differential with regard to quality and area. Thus, taste tests have been developed that use stimuli representing each of the four taste qualities and that test on localized areas of the tongue and palate.

Genetic differences in taste ability can now be distinguished from pathological differences. It is now known, for example, that about one-third of the population are "taste blind" to PTC (phenylthiocarbamide) and its chemical relative PROP (6-n-propylthiouracil). This taste blindness is a simple Mendelian recessive characteristic. "Nontasters" (those who are taste blind) are also insensitive to some other bitter and sweet compounds. For example, nontasters perceive caffeine to be less bitter and sucrose to be less sweet.

Perceptible olfactory losses have proved to be more common than perceptible taste losses. For this reason, the patients that self-refer to taste and smell clinics usually have olfactory losses. The primary etiologies associated with olfactory loss are head trauma, viral infection, and allergy. Interestingly enough, the head trauma and viral infection patients also tend to show localized taste losses of which they are unaware. This surprising inability of patients to report localized taste losses appears to result from a compensatory mechanism. When one area of the tongue is damaged, other areas actually produce sensations that are more intense. This maintains a relative constancy of taste experience.

The clinics have led to growing appreciation of the devastating effects of dysgeusia and parosmia (chronic bad taste and smell sensations that occur in the absence of obvious stimulation). Improved diagnostic procedures have led to the conclusion that many cases are local in origin. That is, the bad tastes and smells are produced by substances actually in the oral or nasal cavities, such as postnasal drip. Recognition of their origin permits rational therapies.

During the last decade, remarkable progress has been made in establishing the nature of the changes that occur in the chemical senses with age. It is now known that age takes a much greater toll on olfaction than on taste. This insight will permit the development of food products tailored for the elderly as well as other individuals with reduced olfactory abilities. The odorant intensities in foods can be artificially intensified without compromising health. The discovery that age takes an especially heavy toll on the ability to smell the mercaptans added as a warning agent to natural gas permits the search for an additive than can be perceived by the elderly.

Program Goals

The program goals for chemosensory sciences can be divided into basic biological and clinical areas.

In the biological area, highest priorities should be assigned to studies of:

- Plasticity, including development, regeneration, and aging.
- Access of infectious agents and toxins to the brain through the olfactory nerve.
- Receptor mechanisms and signal processing throughout central systems.

In the clinical area, highest priorities should be assigned to studies of:

- Epidemiology of taste and smell disorders.
- Development of new diagnostic tests.
- Affective dimensions of taste and smell.
- Associations between chemosensory disorders and altered food intake in aging as well as in various chronic illnesses.
- Effects of environmental agents on chemosensory function.
- Clinical trials to evaluate potential treatment protocols and rehabilitative strategies.

Basic Biological Area

Plasticity, Including Development, Regeneration, and Aging

The olfactory system offers a special opportunity for understanding fundamental mechanisms of neural regeneration, brain development, and plasticity. Olfactory nerve cells (which, in the same cell, include both the receptor ending and the primary fiber) have the unique capacity to grow and divide throughout life. They not only replace themselves (in a few months at most) following traumatic lesions, but they also show a remarkable naturally occurring turnover rate. These are the only known neurons with this property. The capacity of these neurons for continuous replacement outside their normal environments makes them an excellent candidate for experimental procedures involving repair of brain and spinal cord regions. Thus, research to test whether they can serve as a natural cellular substrate for repairing damaged regions within the central nervous system is a high priority.

As with olfaction, taste cells have special properties that contribute to plasticity in the gustatory system. Taste cells renew themselves throughout life in a process of continuous turnover that necessitates remodeling of connections with the afferent taste nerve fibers. The continued renewal of taste receptor cells presumably underlies some of the changing responses, preferences, and aversions for specific foods and flavors observed in the gustatory system from birth through old age.

Taste buds degenerate if the peripheral innervation is cut, and they regenerate upon reinnervation. Taste buds are also susceptible to cycles of degeneration and regeneration associated with treatments such as radiation and chemotherapy. Thus, the taste system offers unique and as yet underutilized opportunities for study of receptor systems that are renewed throughout life. Investigations of basic mechanisms that underlie taste cell turnover and regenerative properties should be pursued and compared with analogous properties in other senses. Development of tissue culture systems will contribute to knowledge about these mechanisms as well as provide models for basic biochemical and electrophysiological studies of taste reception.

Access to the Brain Through the Olfactory Nerve

There is evidence that the olfactory nerve is a route that allows substances from the external world to enter the brain directly, bypassing the blood-brain barrier. That is, chemicals and pathogenic organisms can gain entrance to the central nervous system through the olfactory nerves. It is necessary to determine which of the viruses responsible for CNS infections travel this route and how to block this process. One of the first signs of Alzheimer's disease is loss of olfactory sensation. Does the causative agent use an olfactory entry route and, if so, what might it be? Also, are changes in olfactory sensitivity reliable early diagnostic signs of exposure to environmental toxins, and can the olfactory nerve serve as a route

of entry for pharmacological agents that are excluded from other routes of entry to the central nervous system?

Receptor Mechanisms and Signal Processing Throughout Central Systems

Taste buds and olfactory neurons are located in a key position at the interface between the external sensory world and the internal milieu of the organism. Mechanisms to transduce chemical stimuli into neural signals include diverse processes of transport through ion channels and binding to specific receptor proteins. Diversity in these mechanisms reflects the variety of chemosensory stimuli. Within the central nervous system, various biologic processes of taste and smell are manifest in complex networks that include local circuits and close associations with a variety of areas in the brain. Investigations that use single-cell isolation and recording techniques, biochemical and biophysical techniques, specific genetic strains of experimental animals, molecular and genetic probes, and immunocytochemical techniques are essential for discovering the basic biology of information processing from receptor to the central nervous system. This basic information forms a context for understanding clinically abnormal chemosensory function.

Clinical Area

Epidemiology of Taste and Smell Disorders

There is a paramount need to assess the incidence, prevalence, and risk factors of smell and taste disorders in the U.S. population. A major national study is required to develop these data, with special attention to etiology, geographic and occupational factors, aging, gender differences, environmental exposure, and genetic and socioeconomic factors.

Development of New Diagnostic Techniques

A major problem in the characterization of chemosensory dysfunction is a lack of definitive diagnostic techniques. Psychophysical tests that not only identify who has a chemosensory loss or disability but also indicate the probable cause of that loss or disability are critically needed. In particular, procedures should be developed that more adequately characterize phantom odors and tastes. In conjunction with chemosensory testing, tactile sensitivity measurements in the oral and nasal cavities should prove valuable. One new technique worthy of special pursuit is to biopsy the olfactory mucosa and tongue as a means of looking for possible pathological correlates of particular olfactory and gustatory disorders.

Affective Dimensions of Taste and Smell

The enormous pleasure derived from chemosensory experience ranges from associations with eating and drinking to associations with sexually important odors. Because the chemosensory experiences are so fundamental, losses of taste and smell profoundly diminish the quality of life in especially disturbing ways. The presence of unpleasant phantoms of taste or smell

produces depression and suffering similar to that experienced by patients with chronic pain. Thus, knowledge is needed of the extent to which taste and smell aversions and preferences are acquired or are innate. A more basic understanding of the pleasure associated with taste and smell could indicate how better to alleviate the suffering associated with chemosensory losses. In addition, it will help in the treatment of conditions such as eating disorders that involve disturbances of chemosensory pleasure.

Associations Between Chemosensory Disorders and Altered Food Intake in Aging and in Chronic Illnesses

Some chronic illnesses are associated with chemosensory symptoms that affect food intake. For example, taste loss and/or a metallic dysgeusia can diminish food intake in uremia. A specific loss in the ability to taste salt has been reported to occur with hypertension. This might be expected to cause increased intake of salt, thereby exacerbating the hypertension.

Effects of Environmental Agents on Chemosensory Function

Environmental agents such as pollutants and pathogens can cause serious damage to chemosensory systems. These systems are in direct contact with the environment and are used to warn and protect against the hazards of such environmental stimuli. There is a need to assess the exposure in polluted areas and in the workplace. This need will grow as the amount of chemical waste and pollution increases.

Clinical Trials for Treatments of Chemosensory Disorders and Rehabilitative Strategies

Treatment strategies for some chemosensory disorders are beginning to emerge. For example, topically applied steroids have shown promise in restoring olfactory function in some patients. Clinical trials of such therapies are needed to assess their potential value.

Scientific Research Opportunities, Strategies, and Priorities

Basic Study of the Chemical Senses

Plasticity, Including Development, Regeneration, and Aging

Olfactory Placode in the Development of the Brain: Strategies for Promoting Regeneration and Reconnection in the Central Nervous System. Evidence gathered over the last century suggests that the development of the olfactory organ is related to, and actually conditions, the development of the brain. For instance, when precursors of the olfactory neurons are removed in the frog, the development

of the brain is impaired. Similarly, administration of large doses of some drugs such as vitamin A specifically damage the olfactory system and also impair brain development. Removal of olfactory precursors at a very early embryonic stage impairs the development of the forebrain. In clinical terms, anencephalia may be produced by a primary dysfunction in the development of the olfactory system.

Many conditions of abnormal brain development in humans may be traced to biochemical deficits of nasal origin. This hypothesis is strengthened by the observation that humans born with certain CNS anomalies may also have olfactory deficits.

Human teratology studies of the nose and of the nose-brain relationship from clinical, experimental, and biochemical points of view would be valuable. The regenerative capacity of the olfactory nerve and the actual transplantation of the precursors of the entire nose have allowed construction of super brains in animal models. These experimental findings may prove clinically useful.

The olfactory neuron is the only known nerve cell in vertebrates, including primates, that can be replaced or grow back throughout the full life span. In addition, this exceptional neuron can grow its axon process not only to the regions of the brain related to the processing of olfactory information but also to non-olfactory regions. Conceivably, these new neurons could help the damaged nervous system repair itself following events such as stroke, degenerative syndromes of the central nervous system, accidental lesions of the brain, and spinal cord injury. New techniques of molecular biology and gene transfer may be adapted to induce these neurons to replicate in specific CNS environments, where they can substitute for damaged neurons.

Researchers are learning how to grow olfactory neurons in vitro. Perhaps future neurosurgeons may be able to use such "reserve neurons" in transplant procedures, replacing damaged tissue. Just as kidneys and hearts are now being transplanted, nervous tissue may be transplantable into the central nervous system.

Cloning of Neurons in Culture: Molecular Genetic Techniques. The unique mitotic capacity of the basal cells from which olfactory receptor cells derive permits them to be continuously replaced throughout life. The genetic factors and the control processes that provide for orderly neuron replacement need to be identified. Developing functionally competent clonal cell lines (an effort now under way) would allow investigation of the molecular processes in neuron development, maturation, and aging. Techniques of molecular genetics, including gene splicing, in situ hybridization, and isolation of genetic messages, could then help delineate the processes that regulate neuron differentiation and mitotic inhibition. Related studies of the capacity of neuron stem cells to provide new neural tissue for brain transplant therapy are needed.

The trophic influence of the olfactory receptor neuron on its targets in the brain provides an excellent model of developmental learning and plasticity. When young rats are exposed to the smells of their mother, profound anatomical, neurophysiological, and behavioral changes occur

in the olfactory brain. These effects can be blocked by a specific receptor antagonist (N-methyl-D-aspartate).

Future research opportunities include those to:

- Investigate the teratology of the human nose.
- Identify olfactory factors that control development of the brain.
- Determine the extent to which olfactory tissue can be used to stimulate central nervous system regeneration.
- Apply techniques of molecular genetics to learn whether the olfactory regenerative capacity can be transferred.
- Investigate the olfactory system to facilitate identification of the neurotransmitters involved in learning.

Development of the Taste System. During development of the taste bud, gustatory nerve fibers innervate papillae that have begun to form on the tongue and subsequently penetrate the basement membrane to induce differentiation of cells in the taste bud. Although the development of cell types is apparently sequential, there is little information, beyond an outline of major events, to define interactions among nerve, mesenchyme, and epithelium that organize and maintain gustatory papillae and taste buds.

The role of the papillae in directing early taste fibers, for instance, is unknown. There is no information at a molecular level of possible roles for cell surface or extracellular matrix molecules in establishing the peripheral taste system. At a cellular level, there are no details about numbers of ganglion cells and fibers throughout development, the qualitative and quantitative appearance of cell types in the taste bud, the initiation of the cell turnover cycle, and other major events. In addition, the origin of different taste cell types is not understood. Monoclonal antibodies and other molecular probes may provide markers for cells at specific developmental stages. Cell and tissue culture techniques should be used to study molecular events in taste bud development. Quantitative light and electron microscopy should be applied more extensively in the study of cellular events during development.

Taste bud cells in adults are normally lost and replaced about every 10 days. In experiments, taste buds degenerate after section of innervating taste nerves and subsequently reform during the process of reinnervation by regenerating fibers. In both processes, synapses with afferent fibers that innervate the taste buds must be continually remodeled. This observation leads to important questions about how taste information is coded during this cycle.

Because cells turn over in adult organisms, they readily degenerate (and regenerate) following trauma and certain clinical treatments, such as radiation and chemotherapy. These cycles of degeneration and regeneration can result in the transient loss of smell and taste. When associated with medical treatment, such difficulties interact with other clinical problems and affect a person's appetite.

The technique of growing severed nerves through multi-electrode arrays for chronic recordings in animals provides an important opportunity to study regeneration of taste function after trauma or treatment.

Future research opportunities include those to:

- Use molecular probes as markers for specific taste bud cell types and developmental stages.
- Apply cell and tissue culture techniques to specify development of taste bud cell types and innervation properties.
- Study regenerating gustatory nerves after trauma or various clinical treatments.
- Explore similarities and differences in development and maintenance of olfactory neurons, taste bud cells, and auditory hair cells.

Aging. Although declines in chemosensory acuity are associated with old age, their biological basis is not understood. Olfactory neurons are lost during aging in some species, whereas numbers of taste buds apparently are maintained in humans, nonhuman primates, and rodents. Furthermore, neural taste responses indicate robust gustatory function in old rodents. Also, although olfactory receptor cells decline in numbers, new neurons continue to be produced in old rodents. There is no information, however, about chemosensory receptor changes at biochemical, membrane, or cellular levels, and there are no more than rudimentary data about age-associated differences in central gustatory and olfactory systems. Fundamental neurobiological studies are essential as a basis for understanding taste and smell sensory problems in the elderly.

Future research opportunities include those to:

- Explore the phenomenon of senescence in olfactory and taste systems to understand age-related chemosensory deficits.

Access of Infectious Agents and Toxins to the Brain Through the Olfactory Nerve

Olfactory receptor neurons provide a direct connection between the external environment and the brain. A vigorous transport system moves substances taken up by olfactory nerve cell bodies located in the nasal cavity. The system takes the substances along the nerve into the olfactory bulb and, in some cases, from the bulb into other parts of the central nervous system. A variety of low molecular weight organic chemicals, some proteins, colloidal gold, and pathogens apparently reach the brain by this route. The system potentially can provide a route for administration of therapeutic substances to specific targets in the brain, avoiding the blood-brain barrier, which ordinarily prevents such access.

The olfactory nerve can also serve as a pathway for some toxins and pathogens. Herpes simplex virus type I, some neurotropic arboviruses, and vesicular stomatitis virus, for instance, are transported into the brain through this pathway, where they can spread extensively. According to some studies, the olfactory sensory deficits expressed in early stages of Alzheimer's disease may be the result of a causative agent that gained entry through these receptor neurons.

Studies of this process may have important implications for public health and for improved neuropharmaceuticals.

Future research opportunities include those to:

- Investigate the role of the olfactory nerve in transport of toxins and pathogens to the brain.
- Study the potential of the olfactory nerve for administration of pharmacologically relevant substances to the brain.
- Develop animal models to investigate these processes.

Receptor Mechanisms and Signal Processing

Stimulus Access. To stimulate olfactory receptors, odorant molecules must be transported by airflow to the olfactory cleft where they settle on the surface of the mucosa and then diffuse through the mucus to the receptor cells. Although these processes are not well understood, they could be the basis for some olfactory dysfunctions. The principles of fluid mechanics and mass transfer should be applied to studies of the transport of odorant molecules to receptor cells. Particular attention should be given to how changes in nasal morphology, sniffing characteristics, and secretory processes might alter this transport.

In the gustatory system, tongue and masticatory movements bring stimuli into contact with taste buds in various oral regions. Salivary secretions, stimulated by taste, provide the microenvironment in which stimulus-receptor contact is initiated. Knowledge of taste-salivary interactions is important to understand mechanisms of taste transduction and potential taste problems associated with altered salivary secretion.

Future research opportunities include those to:

- Describe the access of the odorant and tastant molecules to the receptor cells, and investigate the processes that could interfere with this access.
- Understand roles for mucus and saliva in stimulus-receptor interactions.

Chemosensory Transduction. Recent experiments have revealed some of the transduction mechanisms in taste receptor cells. Taste receptor specificity has been shown to be a property of the peripheral taste epithelium. Voltage-sensitive potassium channels that mediate acid-sensation, and amiloride-sensitive sodium channels that mediate salt sensation are located on the cell apex. Specific receptor proteins and a cyclic AMP-dependent phosphorylation of basal potassium channels appear to be involved in sugar-stimulus transduction in some species. Receptor proteins that are coupled to second messenger generating systems or closely associated ion channels have been described in another species.

Genetic methods should be used to identify taste receptor molecules and to dissect the molecular events underlying the sensing process. Meanwhile, assessing the differences among receptor distributions on the cells of the taste bud should elucidate how stimulus discrimination occurs. Little is known about the synaptic relationships and transmitter chemistry connecting receptor cells to afferent nerves or about how message constancy is maintained in the presence of receptor cell turnover. New

techniques involving tissue slice preparations and voltage-sensitive dyes may make the needed studies feasible.

Olfactory stimulus reception and transduction also appear to be complex. Some odorants and guanosine triphosphate stimulate adenylate cyclase activity in preparations of cilia, which are the transduction sites of olfactory receptor neurons. An extracellular protein in mucus binds odorants and may be a component of a stimulus transport system or an odorant-protein stimulus complex. Nonspecific effects of odorants on membrane charge distribution are also known. The stimulus-receptor molecular complex in the membrane remains hypothetical in primates, although its existence in aquatic species has been demonstrated. Although specific anosmias could provide clues to genetically specified receptor molecules, descriptions of such abnormalities are not definitive. A few monoclonal antibodies that mark subsets of receptor neurons with distinctive cell surface properties have been produced, and technology for measuring the differences of response properties among subsets of neurons is slowly emerging.

Prospects for dramatic advances depend upon improved recording methods, such as voltage-sensitive dyes that are effective on small cells. Success in establishing clonal cultures of receptor neuron stem cells or of immortalizing neurons in culture with spliced oncogenes would allow biochemical and genetic analysis of the receptor process.

Future research opportunities include those to:

- Increase application of whole cell and channel recording techniques to augment the description of taste and olfactory transduction processes.
- Pursue techniques to allow characterization and isolation of the receptor/transduction macromolecules.
- Perfect multiple-cell analysis methods, including voltage- and chemical-sensitive dyes and monoclonal antibody markers for olfaction and taste.

By producing animals with specific deficiencies in the perception of odors and tastes, researchers can compare normal animals to genetically incomplete or abnormal animals. The expression of the specific products of every gene (for instance, of a mouse) can provide essential clues for olfactory and gustatory mechanisms and functions. The use of genetic engineering to produce transgenic animals thus provides a powerful technique to study specific chemosensory deficiencies and production of chemical signals at the molecular level.

Future research opportunities include those to:

- Develop clonal preparations of taste and olfactory cell precursors to permit genetic and biochemical analysis of reception and synaptic transmission.
- Develop transgenic animals with specific sensory deficiencies as models for stimulus discrimination and human pathology.

Olfactory and gustatory receptor studies will profit from recently introduced in vitro and animal models. Such methods include purified ciliary membranes that are reconstituted for patch electrode channel analysis, epithelial cell culture and organ culture, epithelial slice preparations, and whole

cilium and whole cell patch recording. Useful animal models for genetic manipulation include *Drosophila* and *Caenorahditis elegans*. A concentrated effort to understand olfactory and gustatory transduction is appropriate.

Coding of stimulus information in receptor cells also is not understood. Questions of spatial organization, receptor-to-central nervous projection organization, and axon-axon interactions, which are central for analysis of the neural message, need experimental analysis. Studies of aquatic and invertebrate species can provide particularly useful information.

Future research opportunities include those to:

- Exploit simple animal and cell models to do combined genetic and physiological analysis of chemoreceptor mechanisms.

Chemotopic Mapping in Olfaction. Different odorants establish different space-time patterns of activity across the olfactory neuroepithelium and olfactory bulb. These different patterns derive from at least two sources: (1) the receptor cells in different regions of the neuroepithelium are differentially sensitive to the same odorants, and (2) different odorant molecules, depending upon their sorption to the neuroepithelium, spread in different space-time patterns across its surface. These space-time patterns of activity could, in analogy to other sensory systems, be one of the bases for olfactory discrimination. If so, maintaining the integrity of these patterns might be necessary for proper olfactory function. Any disruption (such as neuroepithelial disease states and compromises in the normal reconstitution of the receptor cell population) could affect olfactory function. However, at least at the neuroepithelial level, these odorant-dependent patterns of differential activity have not yet been demonstrated in mammals. Despite one paper on a single species, there is no body of behavioral data showing that olfactory discrimination does indeed depend upon these patterns of differential activity. Techniques such as voltage-sensitive dyes are now available to test in mammals whether these possibly all-important patterns of neuroepithelial activity exist and whether they play a role in olfactory discrimination.

Odorant perception involves cranial nerve inputs in addition to input of the olfactory nerve. These inputs include the terminal, trigeminal, glossopharyngeal, and vagus nerves. The contribution of these other neural inputs known, in general, to affect the perception of odorants in humans needs further detailed systematic analysis.

Future research opportunities include those to:

- Test the proposition that patterns of neuroepithelial activity have a role in olfactory discrimination in mammals.
- Determine the detailed contribution of the other cranial nerves, which, in addition to the olfactory nerve, aid in the perception of odorants.

Chemotopic Mapping in Taste. Mechanoreceptors in the skin are arranged in specific anatomical and functional patterns. These patterns of neural connectivity are represented throughout the central nervous system in somatotopic projections that

preserve the peripheral receptor maps. Although the somatotopy forms early in development, these projections subsequently adapt in response to manipulations of the periphery. One such well-studied mechanoreceptor system consists of vibrissae follicles on the rat snout, which are arranged in precise rows. In the taste system, fungiform and circumvallate papillae also are arranged in an orderly pattern, and manipulation of papillae can provide an opportunity to discern whether these arrays of receptor organs project throughout the central nervous system in a somatotopic manner.

Thus far, only a rather coarse somatotopy has been described in the taste system, in which receptor organs in anterior-to-posterior oral pharyngeal locations project in a rostral-to-caudal direction in the brain stem. Knowledge of a possible finer grained somatotopy in the taste system is essential for understanding the basis of localized taste deficits or alterations in sensation. Knowledge of how such a somatotopy is established during development and the extent to which it can be manipulated would contribute to understanding how neural networks are modeled, maintained, and changed over the life cycle.

Basic knowledge of somatotopy is important for understanding organization in the taste system. In addition, knowledge of chemotopy could provide an organizing principle at various neural levels. There is some evidence for coarse gradients of chemotopic representation in the brain stem and for chemotopic representation of various taste qualities in the cortex. Many more studies of chemotopy are needed, however, before any firm conclusions can be drawn. Without knowledge about how the sensory system is organized, the source(s) of taste defects will be elusive.

Future research opportunities include those to:

- Conduct studies to discern whether the central taste system is organized according to somatotopic and chemotopic principles.

Central Olfactory Connections and Transmitter Mechanisms. With techniques of immunohistochemistry and monoclonal antibody labeling, investigators have identified many previously unsuspected connections between the olfactory bulb and much of the rest of the central nervous system. The functions of these newly identified pathways are not fully understood. The central pathways are notable for their large number and their very wide distribution. Olfaction possibly plays major roles in alertness, decision processes, and homeostasis. Similarly, robust efferent paths connect diverse brain regions to the olfactory bulb. The entire system exhibits a surprising degree of plasticity. It changes with sensory deprivation and in normal development over periods longer than those of other parts of the nervous system.

The olfactory bulb has higher concentrations of more varieties of synaptic transmitter receptors than are found in any other part of the brain. Functions of these chemically distinct pathways are imperfectly understood. The identity of the transmitter-mediating neuron receptor to mitral cell signaling is not known, nor is that of the mitral cell output. The catalog of bulb transmitter receptors includes most of the known nervous system transmitters and

modulators. Deciphering the functional organization of this anatomically simple, neurochemically complex part of the brain is a major challenge. Resolution of it will tell much about how the nervous system controls body processes.

The central connections from the olfactory bulbs to the remainder of the brain may constitute numerous parallel processing subsystems. These subsystems may originate in the periphery; at least two morphologically distinct receptor cells can be found in the olfactory epithelium. It is entirely possible that conscious and subconscious processing of olfactory information proceeds simultaneously. However, the kind of information that is processed in each of these subsystems is unknown.

Future research opportunities include those to:

- Characterize central olfactory neurotransmitters and neuromodulators with biochemical, pharmacological, and immunocytochemical techniques.
- Investigate the functional, developmental, and aging roles of the neurochemically distinct olfactory pathways.
- Further examine species differences in central chemosensory neurotransmitters.
- Determine the function of each of the projection pathways from the olfactory bulbs to the remainder of the brain, and decipher the underlying neural code utilized within each pathway.

Organization of Central Taste Pathways and Neurotransmitters. Taste buds are located in three papilla types on the tongue and are on the soft palate, the epiglottis, and the nasoincisor duct in some species. Sensory information from the taste buds projects via branches of three cranial nerves (VII, IX, and X) to the more rostral, or gustatory, end of the nucleus of the solitary tract. In more caudal regions of the solitary nucleus, neuron types are often characterized on the basis of morphology and neurotransmitter properties, many of which are associated with central cardiovascular and respiratory systems. However, there is as yet no comparable information for the gustatory portion of the solitary nucleus. Because the receptor projections to the solitary nucleus are so extensive and via diverse nerves, there may be a microorganization within the nucleus based on cell morphology, transmitter, and functional properties. In other sensory systems, use of Golgi impregnation, intracellular fluorescent markers, and immunological and lectin probes are yielding important data about subsets of neurons.

Taste nerve fibers also provide sensory input for various digestive, ingestive, and protective reflexes including salivation, gastric changes, swallowing, and respiration. The solitary nucleus contains local circuits to connect afferent input to visceral and motoneurons in other parts of the brain stem. The details of these local circuits are poorly understood but can be better described through application of in vitro brain slice preparations, among other techniques.

From the solitary nucleus, taste information is conveyed to the thalamus via one of two routes, depending on the species under consideration: directly from the medulla or via a synapse in the parabrachial nucleus in the pons. From the pons, fiber systems

project to the lateral hypothalamus and basal forebrain areas. To generalize about central gustatory pathways, the extent of species differences in these and other taste projections must be better understood. Similarly, data on cortical taste projections and response characteristics are far from complete. Throughout the central taste system in animals, little is known about neurotransmitters, but such information is accessible with application of neurobiological techniques.

Future research opportunities include those to:

- Describe central projection circuits for taste buds in various oral-pharyngeal locations.
- Further define morphological and functional neuron types in taste pathways.
- Characterize neural transmitters in taste pathways.
- Further examine species differences in central pathways.
- Investigate changes in central circuits associated with early development and aging.

Clinical Study of the Chemical Senses

Epidemiology of Taste and Smell Disorders

Accurate information is lacking on the incidence, prevalence, and risk factors of anosmia, hyposmia, parosmia, ageusia, hypogeusia, and dysgeusia. There is also a need to know the duration of these disorders. This undertaking entails a major national study and should address etiology, geographic and occupational factors, aging, gender differences, environmental exposure, and genetic and socioeconomic factors. Prognosis by etiologic categories should also be addressed.

Etiology of Chemosensory Losses. The three major causes of anosmia and hyposmia are viral upper respiratory infections, head trauma, and nasal-sinus inflammatory disease, including allergic rhinitis and nasal polyposis. Exposure to inhaled toxic chemicals, particularly in the workplace, is also believed to result in a loss of the sense of smell. In addition, there is a well-documented decrement in the sensitivity of the sense of smell with aging. Selected therapeutic regimens (for example, radiotherapy to the head and neck region, and pharmacological agents such as antihypertensive drugs and antibiotics) may also adversely influence chemosensory function.

Taste losses accompany smell losses in some patients with the olfactory etiologies noted above. In addition, taste loss occurs with neurological damage such as Bell's palsy, Ramsey-Hunt syndrome, and some strokes.

Etiology of Dysgeusia and Parosmia. In the chemical senses, patients may perceive sensations, like tinnitus (ringing in the ears) in audition, for which there is no obvious stimulus. Dysgeusia, which is a chronic, usually unpleasant taste that occurs in the absence of obvious stimulation, can originate because there actually is an unidentified stimulus in the mouth producing the taste, such as medication in saliva or two dissimilar metals in tooth fillings that

produce a battery effect, or because the receptor or its central connections are excited, abnormally, by activity in the peripheral nervous system (PNS) or central nervous system. Known CNS causes include epilepsy and tumors. Known PNS causes include pressure on or stretching of the taste nerve.

Parosmia, like dysgeusia, may involve a chronic, usually unpleasant smell that occurs in the absence of obvious stimulation. In addition, in olfaction, one smell may be substituted for another (for example, licorice for mint). These substitutions may be triggered by an odor or may occur spontaneously. Dysgeusia and parosmia can be distinguished by descriptors used by the patient. Dysgeusia involves the use of the four taste quality names (sweet, salty, sour, bitter). Parosmia descriptors may be the names of objects with particular odors (gasoline, garbage), or the patient may be unable to provide any description other than "unpleasant." Parosmia may be associated with conditions of degeneration or regeneration of olfactory tissue. Like dysgeusia, it may also simply reflect perception of a stimulus that is actually present, such as postnasal drip or infectious processes in the nasal cavity. In addition, it may result from activity in the central nervous system. For example, some patients with temporal lobe epilepsy perceive an olfactory aura. Severe parosmia can be such a debilitating disorder that some patients have undergone brain surgery to remove the olfactory bulbs to get relief.

Future research opportunities include those to:

- Determine the incidence, prevalence, and risk factors of anosmia, hyposmia, parosmia, ageusia, hypogeusia, and dysgeusia.
- Determine the course of these disorders over time.

Chemosensation in Systemic Diseases. Chemosensory sensitivity is reduced in certain systemic diseases including hepatic failure, pseudohypoparathyroidism, diabetes, and hypertension. Smell losses are observed during the course of Parkinson's disease and Alzheimer's disease. These losses appear early in the disease processes and do not evidence progressive change. Losses in the sense of smell in Korsakoff's psychosis correlate with the diminution of the major metabolite of norepinephrine in the cerebrospinal fluid. Taste and smell aversion are common in patients with neoplastic diseases, particularly during periods of chemotherapy.

Competent clinical studies are needed to distinguish between taste and smell sense changes. The role of functional G-proteins should be studied. A high priority is the study of the mechanism of chemosensory loss in pseudohypoparathyroidism. Chemosensory losses may provide a simple and noninvasive early means for detecting degenerative neurological diseases, and lead to new insights in etiology and early intervention.

Future research opportunities include those to:

- Find the cause for disorders of chemosensory sensitivity in systemic diseases.
- Determine the prognosis for recovery.

Development of New Diagnostic Tests

Several recently developed psychophysical procedures are currently being used to evaluate taste and smell. These involve thresholds, identification, confusion among stimuli, and suprathreshold scaling of perceived intensities. Taste testing utilizes the whole mouth or can be restricted to particular loci. Olfactory testing can be restricted to one nostril and thus one nasal cavity. In both taste and olfaction, psychophysical tests do not unambiguously relate to etiology.

In olfaction, either new tests or a restructuring of old tests are needed to indicate, for instance, whether the problem is traceable to the central nervous system or the peripheral nervous system, whether it is based upon the olfactory nerve or trigeminal nerve, or whether it is due to a sensorineural defect or airflow occlusion. Such testing might require a more selective choice of odorants—in analogy to tests of color vision. Thus, carefully chosen stimuli might become confused with one another when certain types of underlying malfunctions exist. Alternatively, one can use the tests now available to determine whether patients having the same diagnosis produce the same overall pattern of wrong identifications. Whatever the approach, tests and strategies to identify the different causes for taste and olfactory dysfunctions are needed.

One further concern is whether there are olfactory losses that are specific to particular odorants. Such studies could help address issues about specific anosmias. The results would not only provide important clues with regard to etiology but would also have important implications for a conceptualization of the process underlying olfactory discrimination.

In taste, losses are already known to be specific to taste substances. But can loss be differential within a single quality? For example, can the ability to taste some sweeteners be reduced while others are spared?

Future research opportunities include those to:

- Develop additional diagnostic methods for smell and taste disorders.
- Develop instruments for monitoring the course of these disorders.

Genetic Variation. Because of genetic variation, people do not all live in the same chemosensory worlds. As noted previously in the National Geographic survey, not all individuals smell certain odors. Similarly, all individuals are not equally able to taste certain bitter and sweet compounds. However, relatively few of the many olfactory and taste compounds that may show the effects of genetic variation have been tested.

Genetic variability may underlie some of the variation in preferences for foods and beverages. The potential economic impact is illustrated by consumer acceptance of artificial sweeteners. Aspartame quickly became the artificial sweetener of choice when compared to saccharin. This occurred in part because saccharin has a bitter taste to PTC tasters (those who are heterozygous or homozygous for the dominant gene for tasting PTC), and such tasters make up two-thirds of the population.

Future research opportunities include those to:

- Expand the knowledge about the impact of genetic variation on perceptions of taste and smell.

Olfactory Pathology. The diagnosis and treatment of olfactory impairment has been made difficult by a lack of knowledge of the pathology, especially the neuroepithelial pathology, of olfactory dysfunction. Recently, however, the technology to biopsy safely the olfactory neuroepithelium of humans has become available. It is, therefore, now possible to search for characteristic consistencies in the neuroepithelial pathology of patients having the same olfactory dysfunctions. If such consistencies exist, the diagnosis of future patients could be very much augmented by neuroepithelial biopsies. Furthermore, nasal biopsy will show the extent to which the biology of animal models transfers to humans.

Taste Pathology. Taste papillae can readily be tested for taste sensitivity before they are snipped from the tongue for analytic processing. The technology for this work is now available.

Future research opportunities include those to:

- Relate lingual and mucosal pathologies to smell and taste disorders.

Clinics see patients who in most instances self-refer. Since these patients are predominantly those with olfactory losses, important clinical information about taste is not being collected. Because the taste system has a built-in redundancy, losses often go unnoticed. Thus, to gather accurate taste data, study populations potentially at risk for taste losses are needed. These include groups of patients such as those with Bell's palsy and Ramsey-Hunt syndrome. There are numerous anecdotal accounts of the effects of drugs on taste, yet few controlled clinical studies verify such effects.

Future research opportunities include those to:

- Identify, survey, and study various populations for the loss of the sense of taste.

Animal Models. An approach for identifying the mechanisms basic to a particular olfactory dysfunction is to produce the same dysfunction in animals. For instance, animals now can take the same odorant confusion matrix test as do human patients. By perturbing the animal's olfactory system to produce the same pattern of odorant confusions observed in patients, common mechanisms may be identified.

Future research opportunities include those to:

- Develop animal models for the study of chemosensory deficits.

Affective Dimensions of Taste and Smell

An infant is born liking sweet and disliking bitter, and this affect is relatively stable across the life span, although it may change in degree. In contrast, olfactory affect does not appear to be present at birth and is apparently more labile across the life span. For example, odors paired with beneficial consequences such as calories become pleasant while those paired with illness become unpleasant. The details of this acquired affect require study.

Odors experienced early in life may acquire life-long affect and arouse strong

emotions in adulthood. Loss of the pleasure that has come to be associated with odors and loss of the pleasure associated with taste can seriously impair the quality of life.

Related to the negative affect of chemosensation, some individuals suffer the chronic presence of unpleasant tastes and odors in the absence of obvious stimulation. These sensory phantoms can make eating extremely unpleasant. Research is needed to determine in detail how the diet is altered by these dysfunctions, giving special attention to the nutritional risks engendered.

The affect associated with taste and smell may be disturbed even if taste and smell sensations are not themselves affected. For example, patients with eating disorders (anorexia nervosa or bulimia) show abnormal affect toward food. In addition, aversions to food can be conditioned as noted above. Food with a particular flavor becomes unpleasant when it is followed by nausea even if the nausea is clearly unrelated to the food. Cancer chemotherapy provides a vivid clinical example of conditioned food aversions. Patients experiencing nausea from chemotherapy can develop aversions to foods consumed before the therapy. This is believed to account, in part, for the appetite loss that often accompanies chemotherapy. These aversions can be prevented to some degree by administration of a novel flavor before chemotherapy. The aversion tends to condition the novel flavor and spare the diet. Conditioned food aversions have proven beneficial under some circumstances. Alcoholics have been helped to abstain from the beverages that they abuse by forming conditioned aversions to those beverages.

Studies of animal models suggest that olfactory stimuli play a role in sexual behavior, reproduction, and maternal behavior. Much, if not all, of this behavior may be learned, but the details are lacking. Some studies suggest that these behaviors have unlearned components, and it is possible that human behavior might be similarly controlled. The nature-nurture controversy as well as the extent to which odors are involved in control of social and sexual behavior in reproductive physiology in humans needs elucidation.

Future research opportunities include those to:

- Delineate the innate and acquired factors in chemosensory affect.
- Assess taste and smell affect in clinical testing.
- Determine the role of odors in human social and sexual behavior and reproductive physiology.
- Investigate how smell and taste, in normal and deficient states, impact on food choice and utilization.

Association Between Chemosensory Disorders and Altered Food Intake in Aging and in Various Chronic Illnesses

Role of Chemosenses in Homeostasis: Sodium Responsiveness. The body regulates the concentration of essential electrolytes, including sodium and potassium, by a variety of physiological

mechanisms. A primary stage of control is the initial intake of the electrolyte during feeding, and thus the sense of taste is key in detecting, identifying, and accepting these chemicals.

Important progress has been made in understanding the molecular and cellular aspects of salt taste reception. Converging evidence indicates that a voltage-independent sodium channel, which is blocked by amiloride, mediates at least some components of sodium taste. Developmental studies demonstrate that taste receptors have an early, low sensitivity to sodium and gradually acquire increasing responsiveness to this crucial electrolyte. Other investigations suggest that taste receptors acquire this sodium sensitivity by adding specific membrane components that can be blocked by amiloride. Acquisition of this membrane component apparently can be blocked by dietary salt deprivation during early developmental periods in animal models. Aldosterone or other hormones may play an important role in regulation of the sodium receptor components in the taste bud. If applicable to taste, such hormonal mechanisms would unify regulation of sodium taste receptors with regulation of sodium in the cells throughout the body. Sodium taste receptors might be regulated relatively rapidly in response to diet changes, changes in salivary composition, or plasma electrolyte levels. Thus, the taste system could alter intake appropriately to maintain sodium homeostasis. Understanding the correct regulatory mechanisms will help explain how taste interacts with internal physiological systems to maintain electrolyte and fluid balance in the body. More generally, it is important to study how taste interacts with nutritional states and bodily needs.

Future research opportunities include those to:

- Extend studies of the regulation of salt intake by the chemosensory system.

Aging. Aging takes a greater toll on olfaction than on taste. Since olfaction provides the cues used to identify foods (for instance, chocolate versus vanilla), losses in olfaction diminish the pleasure associated with eating. In addition to the sensory losses sustained in olfaction, there is a cognitive loss. Elderly persons may have difficulty identifying a food even when they can smell it.

There is controversy about whether the losses in olfactory ability are uniform across odorants. This question is not only of basic interest, but it also has many practical implications. Since the odor placed in natural gas to warn consumers of leaks cannot be perceived by many elderly persons, an odorant might be identified that the elderly could detect.

The fact that age spares taste appears to result partially from redundancy in the taste system. The elderly do show losses at some oral loci, but other loci appear to compensate. Nonetheless, the localized losses put elderly subjects at risk for serious consequences if additional localized losses occur. Many elderly persons experience distortions in taste perception. The etiology of such dysfunctions is unknown. The prevalence of taste dysfunctions in the elderly is also unknown. Dysgeusia is often associated with dentures, periodontal disease, and changes in saliva. The exact

mechanism of this decrease, and its incidence and prevalence in this population, are not known. Some studies have shown replacement of areas of olfactory neuroepithelium with respiratory mucous membrane. For example, to what extent are the olfactory changes seen with aging due to pinching of nerve fibers by the cribriform plate? Does the potential of olfactory cells for regeneration change with age?

The relationships between chemosensory ability and food preference, appetite, and satiety are poorly understood. These relationships have implications for the nutritional status of the elderly. What are the best ways to assess olfaction and taste, especially in elderly and demented populations? What is the relationship between psychophysical function and histologic status? How accurate is olfaction a marker for Alzheimer's disease? Knowledge of the histology and distribution of normal and abnormal olfactory neuroepithelium is needed. Is the loss of the sense of smell in aging due to a defect or loss of first-order neurons or to a lesion in the central olfactory pathways? The answer to this question will require the study of olfactory neuroepithelium through biopsy and the study of the olfactory central nervous system in normal subjects of all ages.

Future research opportunities include those to:

- Study the reasons for variability in the chemosensory performance of the growing elderly population.
- Determine whether the lesions responsible for the olfactory loss in the elderly is in the neuroepithelium or the central olfactory pathways.
- Assess the effect of the health status of the elderly on their chemosensory function.

Chronic Illnesses. Some chronic illnesses are associated with chemosensory symptoms that affect food selection and intake. Furthermore, these alterations in diet may compromise the effectiveness of treatments for the disease. Patients with liver disease, for example, report that food no longer tastes good, and they often cease to eat an adequate diet. Chemosensory changes could contribute to these dietary alterations. In smell losses, which can occur with chronic rhinitis, there is some evidence of overeating, possibly due to an attempt to increase chemosensory stimulation. Individuals with diabetes of various forms experience chemosensory deficits, but whether this could result in increased intake of sugar remains to be investigated. Similarly, a specific loss in the ability to taste salt has been reported to occur with hypertension. This might increase salt intake and exacerbate the hypertension.

As another example, hypothyroidism has been associated both with taste losses and with dramatic shifts in body weight. It is unknown whether these are causally linked.

- Investigate how chemosensory disorders impact on food choice in chronic diseases.

Effects of Environmental Agents on Chemosensory Function

Smell and taste receptors are in direct contact with the external environment, and they are at risk of exposure to potentially

harmful infectious or toxic agents. Although these senses alert persons to environmental chemicals such as pollutants, industrial chemicals in the workplace (vapors and dust), and toxic wastes, such chemicals may also damage these vital chemical senses. Such sensory impairment, particularly in the workplace, contributes to employee absenteeism, loss of productivity, and potential litigation. In addition, workers with impaired sensory function are at increased risk (health and safety) of further exposure and harm from deleterious agents. Workers exposed to cadmium, for instance, show a loss of olfactory function.

Future research opportunities include those to:

- Develop detectors to protect specific employee populations, such as fire fighters, police, special hazard teams, and astronauts.
- Pay particular attention to regions of the country where there is a high concentration of industrial pollutants.

Chemical signals transmitted in the environment can exert pronounced physiological effects on primates, ranging from reduced rates of maturation to effective contraception. Humans commonly live in closed microenvironments, such as air conditioned buildings. This kind of environment is magnified in a space capsule, where crew members share high concentrations of body volatiles, which may cause both depression and nausea.

Complaints of environmental odors produced by industrial wastes, municipal disposal systems, and other sources are common. The point at which environmental chemicals become health hazards or cause unreasonable stress is not known. The likelihood that this problem will grow is certain. While an environmental odor can be recognized as unpleasant, the effects on mood, performance, and behavior have had little clinical study. Multidisciplinary research on defining tolerable levels and health effects of environmental chemicals is critically needed.

Future research opportunities include those to:

- Assess the role of pollution and occupational exposure on chemosensory function.
- Protect and monitor the worker for incipient chemosensory losses due to exposure to the occupational environment.

Clinical Trials to Evaluate Potential Treatment Protocols and Rehabilitative Strategies

Prevention of Chemosensory Disorders. The knowledge necessary for prophylaxis of chemosensory loss is largely lacking. There are no specific measures for preventing smell and taste disorders except for individuals to wear gas masks in certain adverse environmental or occupational settings and to avoid contact or inhalation of neurotoxins. Similarly, protective clothing and gloves are effective in preventing tactile disorders from occupational exposures to toxins. In some cases, antibiotic ingestion in the year before onset of a loss of the sense of smell apparently mitigates the severity of the loss. Likewise, administering conjugated estrogens to postmenopausal

women may protect some of them from anosmia and hyposmia. The authenticity of these findings should be studied.

Future research opportunities include those to:

- Develop new means of prevention of chemosensory losses.

Rehabilitation Strategies. The loss of the senses of smell and taste can be categorized as transport disorders, which involve lack of access of stimulus to the receptors, and as sensorineural disorders, which involve damage to the receptors or their central pathways. Transport disorders in smell and taste can be treated rationally with allergy management, antibiotic therapy, and topical and systemic corticosteroid therapy, and with surgery to treat mechanical obstructions, certain kinds of infections, polyps, and tumors. In special cases, such as with laryngectomized patients, training in how to get airflow into the nasal olfactory cleft has proved helpful. A device (the larynx bypass), although somewhat cumbersome for everyday use, has proved very effective in providing laryngectomized patients an ability to smell.

Currently, there are no therapeutic strategies for the sensorineural disorders of smell and taste. Rehabilitative strategies include psychotherapy, devices such as smoke detectors, and the use of flavor enhancers as well as special textures in foods. Future research opportunities include those to:

- Design and implement clinical trials of therapeutic strategies for sensorineural disorders of smell and taste.

See also chapter 7, "Prevention and Rehabilitation."

Research and Training Centers

Research and training centers would help in the more rapid development of diagnostic tests, collection of epidemiologic data, and the application of molecular biology techniques to smell and taste problems.

- Establish NIDCD National Research and Training Centers dedicated to smell and taste to provide research, training, continuing education, and public health information in this neglected field.

See also chapter 7, "National Research and Training Centers."

TOUCH

Overview

The experience of touch is based on the activity of several sensory systems. These systems cooperate with other sensory systems such as taste, the motor system, and the sense of balance to allow identifying objects and properties of objects, manipulating tools, and becoming oriented in space. Touch may also serve as a channel of communication for the blind by means of braille, for the deaf by means of tactile aids, and for the deaf-blind by means of Tadoma. Touch disorders produce an immediate economic impact on those who earn their

living by working with their hands, and they may also be diagnostic of neurological disorders.

Disorders of touch and other bodily sensations can be known or associated with direct trauma to the skin: for example, frostbite, burn, penetrating wounds, or partial or complete amputations of limbs. They are also associated with trauma to the central nervous system, including stroke, penetrating head wounds or concussion, and congenital malformation or perinatal anoxia, and other injuries in gestation or at birth. Estimates of the prevalence of these disorders vary widely, but the number must range from 3 to 5 percent of the population.

In peripheral trauma, the interest of the clinician lies in the problem of restoration of somatosensory function. When plastic reconstructive surgery, for example, is performed on a burn victim or an accident victim whose limb has been restored by microsurgery, an extended period of recovery is inevitable, with attendant rehabilitative efforts involving hours of testing and exercise of the appendage. Much remains to be learned about appropriate tests of function that will yield reliable prognostic indices, but studies in the past 10 years have resulted in the development of several tests of tactual sensitivity. Incorporation of these tests with the recently developed electrophysiological testing of single units of the peripheral nerves (microneurography) in humans may prove to be a powerful tool in the analysis of extent of peripheral damage and the course of recovery.

In adventitious or operative lesions of the central nervous system, tests of somato sensory function serve to delineate the area of damage as well as to track recovery from the insult. In developmental disabilities, sophisticated testing for tactual or haptic skills in infants and children is lagging behind other techniques and deserves much more attention than it has received to date.

During the past decade, advances have been made in the use of the skin as an aid in communication disorders, such as tactual aids for the deaf, blind, and deaf-blind, and in understanding the most effective ways to make use of these devices with the disabled. Multichannel tactual aids, for example, have shown considerable promise in improving lipreading performance. These devices may be particularly useful in cases in which cochlear implants or other surgical procedures are contraindicated. Devices to convert printed characters directly to tactual patterns that can be read by blind persons continue to be improved, as have tactual displays for use in mobility training of blind and deaf-blind persons.

Recent Accomplishments

Recent progress in understanding tactile sensitivity and more general aspects of the sense of touch has resulted from a number of interconnected activities. Measurements of neurophysiological responses of peripheral and central neurons to mechanical stimulation of the skin have benefited psychophysical studies using similar stimuli, and vice versa. Basic studies of tactile sensitivity have guided the development of sensory aids for the disabled, such as

reading aids for the blind. The use of these aids has led to information about some little-known sensory capabilities of the skin, and the aids are being used by scientists to answer basic questions about the sense of touch. The importance of the sense of touch is further discussed elsewhere in this report: tactile aids for the deaf to transduce auditory information, tactile speech production aids, and the role of touch in balance.

The study of tactile sensitivity is also an important topic of investigation in its own right. Because of the interconnectedness of the various areas of investigation in tactile sensitivity, progress in one area can simultaneously open windows in other areas.

Program Goals

The program goals for the study of touch can be divided into basic biological and clinical areas.

Important biological goals include:

- Neurophysiological and psychophysical studies of tactile coding of spatio-temporal patterns.
- Interaction of touch with other senses.

Important clinical goals include:

- Tactile sensory aids including rehabilitation devices for the deaf.
- Testing of tactile sensitivity in the mouth.

Scientific Research Opportunities, Strategies, and Priorities

Basic Study of the Tactile Sense

Tactile Sensory Coding and Cortical Representation

An important technique in the development of a complete understanding of sensory processes has been combined psychophysical-neurophysiological studies. In such studies, psychophysical measurements are obtained, generally from human subjects, and the same stimulus conditions are used to examine neurophysiological responses, typically of monkeys. In an even more intimate conjunction of psychology and physiology, microneurographic studies have examined peripheral nerve fiber responses in awake human subjects who may simultaneously report their sensations. From both repetitive (temporal) stimuli and spatially extended stimuli, considerable information has been generated on how temporal and spatial patterns are encoded for processing by the nervous system.

Building on this foundation, investigators are recording from central projection areas, primarily in somatosensory cortex, to examine how spatial information is encoded and transformed as it moves from primary

projection areas through association areas. Studies of spatial pattern perception with human subjects are providing an understanding of how such patterns are processed and how sequentially presented patterns affect one another. Continuing work in this area offers considerable promise in understanding tactile processing in particular and also in shedding light on problems in other modalities, such as how spatial information is encoded visually. Investigations of somatotopic organization in cortical areas have shown that these areas possess a remarkable degree of plasticity. These results not only suggest an important role for experience in determining cortical organization but also offer insights into possible recovery from peripheral and central neurological injuries. Advances of this kind underscore the importance of parallel measures of neurophysiological and behavioral functioning.

Future research opportunities include those to:

- Investigate psychophysical and neurophysiological processing of spatial/temporal information in the somatosensory system.

Touch and Its Interaction With Other Senses

It is obvious that the human experience of the world is not neatly separated into sensory channels, as so often appears from laboratory investigations. Contact with an object generates pressure sensations often accompanied by either warmth or cold, or possibly itch or pain. One example of such interaction is a temperature-touch illusion in which a thermal stimulus is perceived to be not at the location at which it is presented but at the location of a pressure stimulus appearing elsewhere. Investigations of these other dimensions of touch need to be pursued not only for what they will reveal about the dimensions themselves (for example, studies of cutaneous pain and itch are of obvious importance in therapeutic interventions) but also for what they reveal about interrelations of the encoding mechanisms of the central nervous system.

One obvious place for complex interaction among the senses is in the oral cavity. Appreciation of food is dependent on the combined activities among smell, taste, touch, temperature, and common chemical sensitivity (the "burn" in highly spiced food). Evidence of this kind of interaction is found in the spatial illusion in which the localization of taste is altered by presentation of a tactile stimulus, a counterpart to the temperature illusion noted above. Studies in this area are important for understanding the factors that determine the palatability of food and the detection of patients' sensory deficits as well as for understanding normal interactions among the senses.

Future research opportunities include those to:

- Study interaction among the senses, including touch, temperature, the chemical senses, pain, and itch.

Development of Tactile Stimulators

A major problem in the investigation of tactile sensitivity is the unavailability of tactile stimulators matched to the full range of the skin's sensory capabilities. Such

stimulators are critically important in developing tactile aids for the sensorially disabled, in conducting controlled clinical evaluations, and in gaining a basic understanding of tactile sensitivity. The situation that basic scientists and clinical investigators currently face is similar to that of an audiologist who is without the capability of presenting controlled speech signals. Knowledge has developed to the point that enough is known about tactile sensitivity to specify the parameters of an ideal tactile stimulator, its frequency response, the amplitudes needed, and the spacing between stimulators. The development of such tactile stimulators represents a major technological problem, but its solution would enable basic scientists to investigate the psychophysical and neurophysiological capabilities of the skin in both animal and human subjects, allow the clinician to measure capabilities as well as sensory deficits, and encourage the prosthetics specialist to design devices and procedures that will assist disabled persons.

Future research opportunities include those to:

- Develop new instruments to present controlled, complex, spatiotemporal patterns to the skin.

Haptics

An extremely important function of the sense of touch is in aiding exploration, identification, and manipulation of objects. These activities are part of what is called haptic exploration of the environment. Systematic studies of haptic exploration of objects in normal subjects are important in providing the means to evaluate patients with neural and muscular deficits as well as to follow recovery from reconstructive surgery of the hand.

Providing adequate feedback and control for prosthetic hands and limbs depends upon an understanding of haptics. The need for information about haptics and tactile pattern perception is particularly great at the present time. The field of robotics and teleoperators requires such information in order to produce robotic sensors and manipulators and to feed back information to operators of these devices.

Future research opportunities include those to:

- Investigate object identification and manipulation by the hand.

Clinical Studies of the Tactile Sense

Tactile Sensory Aids

There is a long history of attempts to enlist the aid of the skin to overcome or compensate for blindness and deafness. Braille is perhaps the best known example of using the skin as an alternative channel of communication. More recently, the development of the Optacon has given blind people direct access to reading printed material. The ability of the sense of touch to process speech rapidly is demonstrated by the skill of blind individuals such as Helen Keller in understanding the speaker's vocal effects through their hands. Currently, a number

of tactile aids for speech are being evaluated and used by deaf people (see "Rehabilitation Devices" in chapter 1). In addition, tactile displays are used to receive feedback from artificial hands and limbs and also to control them.

These examples of the use of the skin to replace a sensory channel demonstrate some of the capabilities of the skin in successfully processing complex spatiotemporal patterns. Part of the success is a result of basic work in tactile pattern perception. Interaction between basic and clinical groups will facilitate the prompt transfer of information and hasten the development and improvement of sensory aids.

Future research opportunities include those to:

- Encourage research in tactile sensory aids.
- Foster contact between basic and clinical groups.

Testing of Tactile Sensitivity in the Mouth

In the evaluative procedures for patients with apparent taste or olfactory deficits, a beginning has been made in incorporating tests for tactile sensitivity. Presently, standard magnitude matching procedures involving liquid viscosities or pressure stimuli on the tongue have been included with gustatory or olfactory test batteries. The correlation of chemosensory deficits with alterations in tactile sensitivity may aid diagnosis.

Future research opportunities include those to:

- Develop a greater variety of test devices for assessing tactile sensitivity in the mouth and apply these to a broad patient population.

SUMMARY OF RESEARCH RECOMMENDATIONS: SMELL AND TASTE

Major Scientific Research Opportunities

Plasticity, Including Development, Regeneration, and Aging

- Identify olfactory factors that control development of the brain.
- Use molecular probes as markers for specific taste bud cell types and developmental stages.
- Apply cell and tissue culture techniques to specify development of taste bud cells.
- Explore similarities and differences in development and maintenance of olfactory neurons, taste bud cells, and auditory hair cells.
- Explore the phenomenon of senescence in olfactory and taste systems to understand age-related chemosensory deficits.

Access of Infectious Agents and Toxins to the Brain Through the Olfactory Nerve

- Investigate the role of the olfactory nerve in the transport of toxins and pathogens to the brain.

Receptor Mechanisms and Signal Processing Throughout Central Systems

- Describe the access of odorant and tastant molecules to the receptor cells, including roles for mucus and saliva, and investigate the processes that could interfere with this access.
- Increase application of whole cell and channel recording techniques to augment the description of taste and olfactory transduction processes.
- Develop clonal preparations of taste and olfactory cell precursors to permit genetic and biochemical analysis of reception and synaptic transmission.
- Conduct studies to discern whether the central taste and olfactory systems are organized according to somatotopic and chemotopic principles.
- Characterize central neurotransmitters and neuromodulators with biochemical, pharmacological, and immunocytochemical techniques in olfaction and taste.
- Investigate changes in central taste and olfactory circuits associated with early development and aging.

Major Clinical Research Opportunities

Epidemiology of Smell and Taste Disorders

- Determine the incidence, prevalence, and risk factors of anosmia, hyposmia, parosmia, ageusia, hypogeusia, and dysgeusia, and observe the course of these disorders over time.
- Find the cause for disorders of chemosensory sensitivity in systemic diseases and determine the prognosis for recovery.

Development of New Diagnostic Tests

- Develop additional diagnostic methods for smell and taste disorders as well as instruments for monitoring their course over time.
- Relate lingual and mucosal pathologies to smell and taste disorders.

Affective Dimensions of Taste and Smell

- Delineate the innate and acquired factors in chemosensory affect.
- Determine the role of odors in human social and sexual behavior and reproductive physiology.

- Investigate how smell and taste, in normal and deficient states, impact on food choice and utilization.

Associations Between Chemosensory Disorders and Altered Food Intake in Aging and in Various Chronic Diseases

- Assess the effect of the health status of the elderly on their chemosensory function.
- Investigate how chemosensory disorders impact on food choice in chronic diseases.

Effects of Environmental Agents on Chemosensory Function

- Assess the role of pollution and occupational exposure on chemosensory function.

Clinical Trials to Evaluate Potential Treatment Protocols and Rehabilitative Strategies

- Design and implement clinical trials of therapeutic strategies for sensorineural disorders of smell and taste.

SUMMARY OF RESEARCH RECOMMENDATIONS: TOUCH

Major Scientific Research Opportunities

Tactile Sensory Coding and Central Representation

- Investigate psychophysical and neurophysical processing of spatial/temporal information in the somatosensory system.

Touch and Its Interaction With Other Senses

- Study interaction among the cutaneous senses, including touch, temperature, the chemical senses, pain, and itch, at both psychophysical and neurophysiological levels.
- Study interaction between proprioceptive and cutaneous senses as well as between vision, hearing, and the cutaneous senses.

Major Clinical Research Opportunities

Development of Tactile Stimulators

- Develop more efficient and compact devices to present controlled, complex spatiotemporal patterns to the skin.

The Haptic Exploration of Objects and Surfaces

- Investigate individual differences in abilities for object identification by tactile exploration in normal and patient populations.

Tactile Sensory Aids

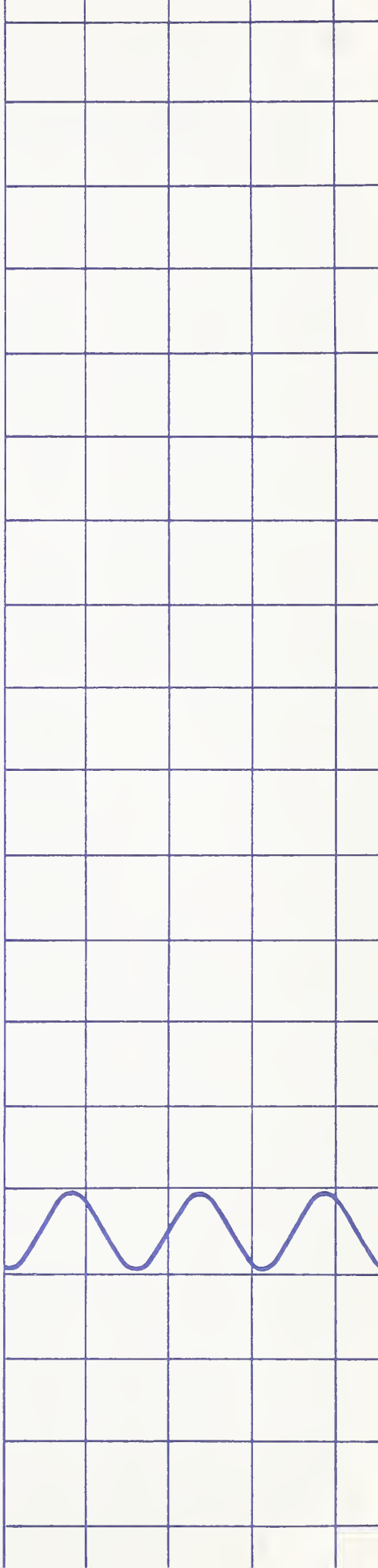
- Encourage research in tactile sensory aids, including speech-analyzing, reading, and mobility aids for deaf and deaf-blind persons.

Testing of Tactile Sensitivity in the Mouth

- Develop devices and techniques for testing tactile sensitivity in the oral-nasal cavity, and apply these to the population of patients with taste and olfactory complaints.

7.

Cross-Cutting Issues



CONTENTS

PREVENTION AND REHABILITATION 231

Prevention 231
Rehabilitation 232
Research Priorities 232

NATIONAL RESEARCH AND TRAINING CENTERS 234

Special Features 234
Benefits 234

DATA SYSTEMS AND POPULATION-BASED STUDIES 235

The Need for Expanded Data Systems 235

Research Strategies and Priorities 238

RESEARCH TRAINING AND RESEARCH PERSONNEL RESOURCES 239

Current Support for Research Personnel Resources and Training 240
Perceived Research Training Needs 240
Past and Current Problems in Recruiting Investigators 242
Strategies for Recruiting Additional Research Personnel 244
Specific Recommendations 245

NATIONAL INFORMATION CLEARINGHOUSE 247

MULTICULTURAL RESEARCH 249

7.
Cross-Cutting Issues

PREVENTION AND REHABILITATION

The six Task Force scientific areas share the twin goals of prevention and rehabilitation. Each area naturally has special needs and concerns in this regard, and they are addressed in the individual chapters. This discussion focuses on topics and principles that have general application to efforts to prevent communication disorders and rehabilitate affected individuals.

Such efforts are typically multidisciplinary. The NIDCD should therefore take the initiative in promoting and funding cooperative and interdisciplinary studies in the area of prevention and rehabilitation. Some of the most promising ideas can come from consumers and members of the public who are currently involved in rehabilitation or habilitation activities. Involvement of this constituency should be encouraged.

Prevention

The term "prevention" encompasses:

- *Primary prevention:* Prevention of an initial disorder where the cause is known. Activities contributing to primary prevention include immunization, control of environmental factors such as noise level and air quality, and genetic counseling.
- *Secondary prevention:* Prevention of progression beyond initial appearance, with possible permanent effects. Activities contributing to secondary prevention include the timely treatment of middle ear infections; identification of balance, taste, and olfaction problems in the elderly; and early attention to developmentally sensitive disorders such as stuttering.
- *Prevention of sequelae:* Early intervention to prevent other consequences that result from the initial disorder. Language problems consequent to congenital hearing impairment, for example, show the wide range and complexity of such sequelae, from specific deficits in language development to more general social and psychological problems created by perceptions and reactions of others to an individual's initial disorder.

Effective prevention programs require:

- An understanding of the mechanisms leading to an impairment.
- Identification of risk factors.
- Identification of populations at risk.
- Availability of screening to identify individuals with impairments.

Education is a key to effective prevention. Education programs should be directed to:

- The public at large.
- Populations known to be at risk (or their parents).
- Physicians, nurses, and other providers of health care and education.

Rehabilitation

The term "rehabilitation" (or "habilitation" in cases of congenital and perinatally acquired disorders) applies to intervention procedures and devices designed to raise an individual's level of function. Rehabilitation focuses on providing the means for disabled people to function independently and to maximize their abilities. It includes:

- *Prosthetic intervention:* Examples include the use of hearing aids, speech-augmenting electronic devices, and devices helping with the hyposmia of laryngectomy to produce corrective nasal airflows.
- *Circumventive intervention:* Examples include the use of alternate sensory modalities, languages, and codes for communication, education, and training.
- *Preventive intervention:* Activities such as training in English language for hearing impaired children focused on preventing adverse effects of hearing loss; or, in the case of chemosensory disorders, simply making individuals aware of their problem so that they can begin to cope.
- *Remedial intervention:* Activities designed to reduce both developmental delays and postdevelopmental losses (for example, adult aphasia).
- *Medical and surgical interventions:* Procedures other than those designed to cure. Examples include the implantation of cochlear electrodes in the management of deafness, and electrical stimulation of paralyzed vocal cords.

Effective rehabilitation programs require:

- An understanding of the nature and mechanisms of the disorder and its interactions with other aspects of development and function.
- An understanding of developmental and learning processes.
- Accurate assessment and description of the impairment, preserved abilities, and any sequelae.
- Capitalization on an individual's strengths rather than an exclusive focus on impairments.
- Availability of knowledgeable and trained personnel.
- Periodic assessment of progress and evaluation of rehabilitation procedures.

Early identification is a key to successful rehabilitation and habilitation.

Research Priorities

To further prevention and rehabilitation in disorders of hearing, balance, voice, speech, language, taste, smell, and touch,

priority should be given to the following types of research:

- Development of vaccines to immunize against infectious diseases, such as meningitis and otitis media, which result in deafness and other communication disorders. This is one of the most direct—and promising—methods of primary prevention.
- Studies of prevention through the control and elimination of environmental agents such as air pollutants and noise.
- Development of educational programs designed to improve the identification of individuals who are affected or at risk, and programs to increase professional awareness of available options for treatment and rehabilitation.
- Development and evaluation of procedures for early identification. This effort includes studies of the application of existing procedures and development of new ones.
- Studies of sensory substitution and integration as they affect the rehabilitation process. This research is especially important since much of rehabilitation involves the co-opting of other senses and sensory aids to take over some of the function lost through disorder or disease.
- Clinical trials for empirical validation of existing interventions and for generation of information leading to improved rehabilitation procedures.
- Studies of successful rehabilitation. Understanding what makes an attempt at rehabilitation successful should lead to the development of programs that offer greater economy and better results.
- Validation and standardization of new methods of assessment, including evaluations of performance and of the outcomes of interventions.
- Research on technological methods of assessing performance of individuals, diagnosis of impairment, quantification of function, determination of need for intervention, and evaluation of the success of intervention.
- Study and development of assistive technology and equipment, including prosthetic devices, designed to promote and to enhance rehabilitation. Efforts should include the prescriptive design of appropriate instrumentation.
- Studies of the overall cost-effectiveness of rehabilitation. The process of rehabilitation is expensive, and it is absolutely essential to keep ethical considerations in full view in any study of the cost-effectiveness of rehabilitation. One device, procedure, or policy may be more "cost-effective" than another when judged only in terms of financial cost, but the question of human costs must be paramount in the evaluation of any manipulation or intervention directed to the rehabilitation of people whose ability to communicate according to societal norms has been compromised.

NATIONAL RESEARCH AND TRAINING CENTERS

Public Law 100-553 mandates the development, modernization, and operation of research centers for studies of deafness and other communication disorders. This mandate permits the development of a unique and flexible funding capability for an integration of multiple research, research training, and dissemination functions under a single mechanism. Taking advantage of this capability, the NIDCD National Research and Training Centers (NRTCs) should be multidisciplinary in approach, and each should have a central, unifying theme, combining basic and clinical research and strongly promoting interactions among scientists across the research spectrum.

Special Features

The centers should typically share common resources, including technical, clerical, and administrative personnel, instrumentation, computer resources, subject populations, and data bases. Importantly distinct from research centers in the past, the NIDCD NRTCs are intended to have a broad training and technology transfer mission, including:

- Cross-disciplinary training of residents, pre- and postdoctoral fellows, visiting scientists, and students.
- Continuing education for physicians, professionals dealing with the hearing, language, and speech impaired individuals, and for other professionals.
- Dissemination of information to the public.
- Transfer of technology for clinical utilization.

In addition and where appropriate, the centers may pursue the following activities:

- Evaluate the effectiveness of existing, new, and improved methods of (1) education and rehabilitation (especially with hearing impaired and language impaired children, the elderly, and multicultural populations), and (2) detection, assessment, and treatment.
- Undertake relevant epidemiologic and genetic studies.
- Expedite transfer of technology for commercial product development.
- Interact with other centers, research groups, and researchers on matters of common technical, administrative, and resource interests.

Benefits

As currently outlined, these centers will substantially enhance research progress in deafness and other communication disorders. They will have direct scientific impact in the following ways:

- Stimulate research in important but neglected clinical and basic areas.
- Focus needed scientific work on particular disorders.
- Capitalize on research developments offering unusual opportunities for advances in knowledge and clinical management.

- Enhance identification of clinical problems that are particularly amenable to basic science research approaches.
- Provide broad-based and thorough solutions to complex human communication problems.
- Accelerate transfer of state-of-the-art technology to patient management.
- Stimulate development of high-quality health care.
- Provide more efficient use of resources not normally available to individual researchers.

Further, these centers should strengthen the quality and increase the number of investigators by providing long-term stability for careers of programmatic research, by virtue of activities that will:

- Recruit scientists from basic and clinical areas.
- Enrich the multidisciplinary training environment for experienced and inexperienced researchers.
- Develop a critical mass of dedicated investigators working together in areas of communication problems.
- Encourage careers in clinical investigation.

Finally, but importantly, these centers also will provide unique information resources that will help to:

- Enhance the identification and guide directions of important basic science areas from clinical observations, and thus broadly influence that basic science.
- Provide for changes in direction of basic science questions based on awareness of clinical data.
- Heighten awareness of communication problems by the larger biomedical and lay communities.
- Inform the public as well as the general health care community regarding important research advances, preventive measures, mechanisms of patient referral, and sources of care.

DATA SYSTEMS AND POPULATION-BASED STUDIES

There are no existing incidence data on communication disorders on a national level. While prevalence data exist, they do not adequately meet the complete needs of the research and clinical communities and consumer groups. Existing data bases are described briefly in Table 1.

The Need for Expanded Data Systems

It is necessary to obtain sufficient data to study the incidence, prevalence, and risk factors for deafness and other communication disorders in the United States. This need applies to deafness and hearing impairment; vestibular disorders; speech, language, and voice disorders; and disorders of touch, taste, and smell. Such studies should span the entire life cycle, from infancy to old age, and should include population samples reflective of the various cultural and ethnic backgrounds of the U.S. population.

TABLE 1

EXISTING SOURCES OF DATA ON COMMUNICATION DISORDERS

Source	Project	Dates	Type of Data
National Center for Health Statistics (NCHS)	National Health Examination Surveys (NHES):		
	NHES-I	1962-64	Audiometry on adults
	NHES-II*	1963-65	Audiometry on children 6-11
	NHES-III*	1966-70	Audiometry on youths 12-17
	National Health and Nutrition Examination Surveys (NHANES):		
	NHANES-I*	1971-75	Audiometry on adults, self-reported hearing loss
	NHANES-I Epidemiologic Followup*	1982-84	Self-reported hearing loss
	NHANES-II	1976-80	Audiometry on children 4-19;* speech recordings on children 4-6
	Hispanic HANES (HHANES)	1982-84	Audiometry* and tympanometry, ages 6-74
	NHANES-III	1989-95 (in the field)	Audiometry and tympanometry, ages 6-19
	National Health Interview Surveys (NHIS)	Continuous, 1969-87	Self-reported hearing loss
	Hearing Impairment Supplement	1971	
	Hearing Impairment Supplement	1977	
	National Ambulatory Medical Care Survey (NAMCS)	1973, 1975-81, 1985	Physician visits, drug mentions

TABLE 1 (continued)

EXISTING SOURCES OF DATA ON COMMUNICATION DISORDERS

Source	Project	Dates	Type of Data
	National Nursing Home Survey (NNHS)	1963-69, 1973-74, 1977, 1985	Staff-reported impairments
National Institute of Neurological Disorders and Stroke (NINDS)	National Collaborative Perinatal Project (NCPP)	1958-65, with 7-year followup	Pregnancy data, perinatal data, speech samples of children at 7 years of age
National Heart, Lung, and Blood Institute (NHLBI)	Framingham Heart Study	1948-90	Detailed physical exams, audiometry on adults 57-89 years of age
Bureau of the Census	Survey of Income and Program Participation	1984	Self-reported hearing loss
Gallaudet University	Annual Survey of Hearing Impaired Children and Youth	Annually since 1968	Severely hearing impaired, 0-21 years of age
United Nations Statistical Office	United Nations Disability Statistics Data Base	1987	Various disabilities in 75 countries
State and local studies	E.g., New Jersey, Michigan, Utah, East Boston, New Haven (Connecticut), Iowa	Various	Various, primarily hearing impairment
International studies	United Kingdom: Institute of Hearing Research/Medical Research Council	1980-82	Audiometry, tinnitus in British adults
	Sweden: University of Gothenburg	1971-72, 5-year followup	Audiometry, 70-year-old men and women

*Public use data tape is available from the National Technical Information Service (NTIS)

It is important that these studies address testable hypotheses, especially in regard to risk and other factors, such as onset, complications, and severity.

In regard to hearing impairment and deafness, attention should be given to otitis media, sudden hearing loss, age-related hearing loss, congenital/inherited hearingloss, and noise-induced hearing loss. Factors associated with hearing loss such as family history, age, and medical history should be documented. Resources should be committed to genetic studies as well. Serum and lymphocyte samples should be obtained and stored for later analysis as new methods and technologies evolve.

In regard to language and speech, there is an additional need to obtain normative data about a variety of speech, language, and voice behaviors.

In regard to vestibular disorders, there is a need for epidemiologic data, with emphasis on possible etiological factors, and for comprehensive clinical data, including quantitative test results.

In regard to the chemical senses, epidemiologic studies must be especially sensitive to regional differences and social and ethnic differences.

Special consideration should be given to maintaining and upgrading temporal bone banks, particularly in regard to normal data on the peripheral auditory and vestibular systems. In addition, attention should be given to the acquisition and analysis of tissue from the central nervous system, serum, and lymphocytes. Great advances should be made when contemporary diagnostic methods are used premorbidly and when modern research tools are applied to the materials.

Research Strategies and Priorities

The Task Force is virtually unanimous in its support for epidemiologic studies of deafness and other communication disorders. In light of the magnitude of the studies proposed in this report and the complex nature of these undertakings, there is a clear need to establish an Epidemiology Division within the NIDCD.

This division would serve as the Federal focus for epidemiologic and public health concerns relevant to the NIDCD. It is recognized that the operation of the division will be an expensive undertaking, but it is necessary because of the importance of epidemiologic study to the mission of the NIDCD.

A first priority should be the formulation of an advisory committee for the Epidemiology Division. Its composition should include epidemiologic experts, appropriate clinicians and scientists, individuals from the deaf community, persons who represent the communicatively impaired, persons from industry and government, and representatives of multicultural populations. With representatives from all these areas, the unique needs of each should be met.

The agenda of the advisory committee should include the following:

- Improved utilization of current data bases and needed modifications, including research training opportunities.
- Consideration of scientific and professional issues, including terminology and operational definitions of deafness and other communication disorders, and the development of test instruments.

- Establishment of a communication disorders registry.
- Strategic planning with respect to priorities, cost:benefit of epidemiologic investigations, and the allocation of resources.
- Surveys of the health status and needs of the deaf population.

The advisory committee should also consider mechanisms appropriate to achieve interaction with other agencies, such as the National Center for Health Statistics and with the NIDCD National Information Clearinghouse. (See below.)

RESEARCH TRAINING AND RESEARCH PERSONNEL RESOURCES

The current personnel needs for research on deafness and other communication disorders is a primary concern of the Task Force assembled to develop this National Strategic Research Plan of the NIDCD. Many of the new institute's program areas are now sustained only by small groups of active investigators. Important contemporary scientific disciplines including genetics, immunology, cell and molecular biology, biochemistry, and epidemiology are lacking or are seriously underrepresented in many NIDCD areas. These groups are threatened with extinction as the present leadership approaches retirement. In addition, clinically trained M.D.'s and Ph.D.'s presently interacting with the basic scientists are extremely rare and essentially nonexistent in some program areas.

The complexity of modern biomedical research and the subsequent transfer of research results into clinical practice and public behavior demands long periods of research training in an increasing number of discrete areas of specialization. In addition, the pace at which biomedical research is now progressing may quickly render an investigator's acquired knowledge and skills insufficient to address a problem with the most appropriate combinations of disciplinary skills. Investigators must update their knowledge and skills on a continuing basis both inside and outside their own disciplines. Consequently, cross-disciplinary research training should become a rule rather than an exception. To date, mechanisms designed to resolve these problems have been few.

A first priority of the new institute should be to address this critical shortage of research personnel and training support. The dimensions of the problem should be carefully addressed, both qualitatively and quantitatively. Only with such data at hand can the institute respond to pressing needs specifically and quickly, in a cost-effective manner. It is imperative to develop an active, long-range master plan for fostering a cohesive, disciplinarily diverse community of investigators dedicated to advancing research on human communication and its disorders.

A continuation of the current crisis in deafness and communication disorders

research training could have devastating effects on the NIDCD's research program well into the next century. In some research institutions, current deficiencies in the support and retention of scientists dedicated to the study of deafness and other communication disorders may already be almost insurmountable.

The discussion below, of necessity, is based on incomplete information. It is a preliminary review of personnel resources and needs across all disciplines required for research on deafness and other communication disorders.

Current Support for Research Personnel Resources and Training

Available data provide preliminary information for an accurate analysis of either active or developing research personnel resources for biomedical research on deafness and other communication disorders.

The number of training awards transferred from the National Institute of Neurological and Communicative Disorders and Stroke (NINCDS) suggests that slightly more than 60 percent of trainees are in various fields of basic research and slightly more than 40 percent are in clinical research areas. This estimate is based on all extant training mechanisms: individual and institutional National Research Service Awards (NRSAs) and the several types of individual career development award programs.

The Task Force recommended that an analysis of current NIDCD research trainees across all NRSA and career development programs be conducted at NIH. The results of that analysis are in Table 2.

The existing training grants and awards were also analyzed to delineate the nature of the training programs and to assess the distribution of positions across disciplines. The NIDCD should update these data periodically and analyze trends.

Perceived Research Training Needs

Affecting all research areas:

- There is an urgent need to determine the research personnel needs in all areas affecting hearing, balance, voice, speech, language, swallowing, smell, taste, and touch.
- There is a critical need to recruit and train physician investigators, especially otolaryngologists, to interface with the basic researchers in the fields of hearing, balance, voice, speech, language, swallowing, smell, taste, and touch.
- The demand for basic researchers trained in the rapidly advancing fields of science such as biochemistry, cell and molecular biology, molecular genetics, and bioengineering exceeds the supply. This pool of expertise is essential to sustain the rapidly evolving research on the biology of normal and abnormal human communication and deafness and, where appropriate, to devise new strategies for treatment and rehabilitation.

TABLE 2

ANALYSIS OF CURRENT NIDCD RESEARCH TRAINEES (FTTP), MARCH, 1989

Program Area	KO4		KO7/8		F32 (33)		T32		T32	Total
	Ph.D.	M.D.	Ph.D.	M.D.	Ph.D.	M.D.	Ph.D.	M.D.	Pre-doctoral	
Hearing	4		4	2	11(1)	2	19	7	8	58
Otolaryngology				4			3	14	.5	21.5
Balance			2	2			1			5
Speech	1		2	4	1		6		3	17
Language	4			1	3		5	1		14
Taste	2				2		6		2	12
Smell	1		1	1	2(1)					6
Subtotal	12	0	9	14	21	2	40	22	13.5	
Total	12		23		23		62		13.5	133.5

FTTP = Full time training position
KO4 = Research Career Development Awards
KO7/8 = Teacher/Clinical Investigator Awards
F32(33) = Individual NRSAs (Senior Individual NRSAs)
T32 = Institutional NRSAs

Disciplines of NIDCD Trainees by Degree
(P = Ph.D.; M = M.D.)

Hearing: physiology (13P, 1M), anatomy (4P, 4M), neuroscience (4P, 2M), biochemistry (2P, 2M) immunology (2M), audiology (5P), psychoacoustics (6P), developmental neurobiology (1P), biomedical engineering (1P), bioacoustics (1P), auditory perception (1P), psychology (1P). Predoctoral: 3 physiology, 2 neuroscience, 1 biomedical engineering, 2 bioacoustics.

Otolaryngology: general (2P, 7M), pediatric (4M), neurotology (1P, 3M), laryngology (3M), histopathology (1M). Predoctoral: 1/2 general.

Balance: vestibular physiology (1P, 2M), vestibular anatomy (2P). No predoctoral.

Speech: physiology (1P, 3M), communication (2P), histology (1M), communications engineering (2P), development (1P), science (3P), perception (1P). Predoctoral: 1 communications engineering, 2 speech science.

Language: linguistic science (3P), psycholinguistics (2P), neurolinguistics (2P), dyslexia (1M), neuropsychology (2P, 1M), neurobiology (1P), experimental phonology (1P), developmental psychology (1P). No predoctoral.

Taste: sensory biology (3P), neurochemistry (1P), sensory psychology (3P), physiology (1P), anatomy (1P), nutrition (1P). Predoctoral: 2 sensory biology.
Smell: molecular biology (2P, 1M), physiology (2P), biophysics (1P). No predoctoral.

- There is a critical need to train pathologists with expertise in temporal bone pathology and other human tissues and organ systems related to deafness, communication, balance, and chemosensory disorders.
- There is a critical need to support short-term research training in new technologies such as tissue culture, molecular biology, molecular genetics, hybridoma techniques, imaging modalities, and computer applications. This training is needed for both established and new investigators.
- There is a need to initiate additional training programs designed to attract junior investigators (undergraduate, predoctoral, doctoral, medical student, and resident levels) to research careers related to communication disorders. The lack of ethnic minority investigators in all disciplines is of particular concern, as is the absence of investigators who are deaf or who have communication disorders.

Affecting specific program areas:

- There is a need to recruit large numbers of high-quality graduate students in the field of auditory research. This need is particularly urgent because of the recent decline in both the number and quality of students entering graduate training.
- There is an urgent need to recruit and train researchers in the multidisciplinary fields of science relating to balance disorders. The shortage of clinical investigators active in this area is especially noteworthy.
- To bridge the widening gap between the rapid development of basic research and clinical application, there is a need to initiate multi- and interdisciplinary training in the fields of speech and language.
- At universities, voice science and pathology have frequently not kept close ties with speech and voice training. Interdisciplinary training of individuals in both areas could bring these varied approaches to bear on present and future voice research.
- There is a critical need to recruit and train basic researchers (for example, psychophysicists) and physicians in the chemosensory field, to establish clinical research training programs in chemosensory disorders, and to initiate multi- and interdisciplinary research training in smell and taste. This is particularly critical for otolaryngologists because of the significant decline in those engaged in research careers in recent years. Basic research in smell and taste has advanced rapidly in the last decade, but public demand for clinical care cannot be met without the kind of clinical applied research most likely to be undertaken by scientific investigators.

Past and Current Problems in Recruiting Investigators

- The lack of training funds, an uncertainty about acquiring training grants, the decrease in pre- and postdoctoral

positions, and apprehensions about the future of research grant funding have all contributed to a significant decline in the number of trainees and of the pool of postdoctoral candidates. The consequence of uncertainty in funding is particularly severe for interdisciplinary areas of research, since they are unlikely to be supported by their academic institutions when there is an interruption of training funds.

- The incentive for research training of otolaryngologists and other clinicians in audiology and speech-language pathology has been eroded in recent years by greater demands on all clinicians to generate revenue, often including all of their own salaries. This situation is worsening. In particular, stipends for physicians undertaking research training after completing their clinical training do not begin to compensate for their department's loss of clinical income as a result of research training activities. A supportive chairperson is crucial. Support for basic science research in clinical departments and cross-appointments between basic science and clinical departments must be fostered by department chairpersons and university administrators.
- Many of the disciplines within communication disorders research are relatively new, and individual laboratories are small and widely scattered. In research areas such as disorders of language, taste, smell, and balance, there is very little, if any, coordinated postdoctoral training of M.D.'s or Ph.D.'s.
- Underfunding of training over the years has created critical shortages of trained research personnel. As noted, there are now approximately 100 to 110 individuals in training for future research on deafness and other communication disorders. This number represents between 17 and 18 percent of the 581 trainees being supported by the NINCDS prior to the creation of the NIDCD. The approximately 108 NRSA trainees within the new institute represent less than one percent of the 10,000 to 11,000 trainees supported annually by the NRSA program.
- While the NRSA appropriation for all of the NIH has increased in current dollars, the purchasing power of the appropriations in terms of constant dollars has declined from $131 million in 1974 to $115 million in 1989. The recent increases in trainee stipends, in combination with the devaluation of training support dollars during the past decades, aggravate the crisis situation. The resulting contraction of purchasing power of NRSA training dollars is eliminating established research training programs at the very time that deafness and other communication disorders have been revealed to be underinvestigated health problems of major significance.
- All Task Force participants called for more information on the epidemiology of deafness and other communication disorders. This same information need was cited 10 years ago in the 1979 *Report of the Panel on Communicative Disorders* (NIH Publ. No. 79-1914). There is

a general shortage of epidemiologists in the biomedical sciences and little awareness of the demand for information about communication disorders.

- With rapid developments occurring in basic science areas related to deafness and other communication disorders, a large gap is occurring between the basic sciences and the clinical sciences. New clinical research findings and technologies developed in research centers may not reach the health care delivery system to the benefit of patients. Persons should be trained to develop and improve continuing education programs and other methods of dissemination of information to primary care physicians, audiologists and other professionals, and to patients.

Strategies for Recruiting Additional Research Personnel

- The program directors of academic clinical departments who teach and train personnel for the care of people with communication disorders must be led to recognize their responsibilities for placing a high priority on the research skills and interests of their present and future faculty. Directors should be encouraged to select a significant number of research-oriented trainees for grooming as future faculty and should support trainee and faculty needs for time, space, facilities, and start-up funds essential to research career development.
- Recruitment of students and postdoctoral fellows from basic science disciplines and engineering is an essential strategy to maintain research excellence and state-of-the-art development in the interdisciplinary fields now concerned with deafness and other communication disorders. The well-publicized availability of training funds would be a very effective recruiting tool.
- Financially realistic incentives are required to attract physicians and clinically experienced audiologists and speech-language pathologists into research training. It also is necessary to emphasize clinical research in underrepresented areas such as smell and taste, voice, speech, language, touch, and balance research. In addition, new mechanisms for funding research training for clinicians in other areas may be required. Encouraging basic scientists to obtain exposure to clinical problems would also increase the numbers of well-trained clinical investigators.
- A greater focus on training M.D.'s and Ph.D.'s to participate in well-designed clinical research programs is likely to result in the recruitment of additional physician investigators who wish to focus their research efforts on the clinical portion of the biomedical research spectrum. Clinical research training would be fostered by establishing more clinical research centers featuring clinical researchers as role models. Currently, less than one percent of otolaryngologists and investigators in hearing, speech, and language hold research grants. The percent of clinicians holding research grants in the chemical

senses is equally dismal. This base is dangerously small for the transfer of new knowledge and improved methods of care into actual use with patients.

- It is necessary to foster growth of smaller research disciplines in order to provide a stable environment for training. In particular, in research areas such as language disorders or the relatively new disciplines of balance disorder sciences and chemical senses, little, if any, coordinated postdoctoral training of M.D.'s or Ph.D.'s exists. It is imperative to provide growth and development opportunities in these areas of research, for instance, through attracting established basic researchers and additional new investigators.
- Because it has proven so difficult to persuade epidemiologists to leave well-established fields of health research to pursue careers in communication disorders epidemiology, special incentives must be used to establish and develop this field as an integral part of communications disorders research. Two avenues appear promising: the NIDCD could offer contracts to purchase the needed studies, and through its Division of Epidemiology, it could institute special epidemiology training programs focused on application to deafness and other communication disorders.

Specific Recommendations

- A contract should be let to conduct a comprehensive personnel resources survey that would indicate the number of current research trainees by discipline. The survey should distinguish between basic and clinical research training, or a combination of the two, and project future personnel resource needs in each program area. The ages and disciplinary backgrounds of established investigators also merit attention in this evaluation.
- Training should be a major program in the National Research and Training Centers (see above). The program should span all educational levels.
- To reverse the erosion of funds to support training, drastic and immediate measures are needed. Increased funding of institutional training grants is essential as well as an expansion of other mechanisms.
- Potential trainees are often lost because of overly restrictive position descriptions in a training program. Institutional training grants should be made more flexible and allow for training of predoctoral students, Ph.D.'s, and M.D.'s at the training director's discretion.
- Stipends for physicians, audiologists, and speech and language pathologists undertaking research training should be commensurate with their level of clinical training. Physicians should have the opportunity to pursue at least a two-year period of research training before, during, or after residency training. Flexible programs should be available to accommodate differences in the clinical training programs of medical and surgical specialties.
- The future portfolio of institutional training grants should include grants focused on teaching the methodology of contemporary clinical research as

applied to deafness and other communication disorders.

- A mentor award program should be established. This would enable established but isolated investigators to recruit trainees and provide research training in the absence of formal training grants (T32) to their institutions. Eligible mentors would be holders of research grants (RO1) who would have the option of applying for supplemental funds designated for research training for the duration of their research grants. In view of the interdisciplinary nature of research in most areas of concern and the underutilization of several powerful scientific disciplines, recruitment of mentors from underrepresented basic disciplines is highly desired. The mentor award program should be designed to encourage such needed interactions. This mechanism has the advantage of quickly increasing the number of investigators in training without greatly increasing administrative costs or adding costly mechanisms of evaluation.
- A special fellowship program should be instituted to attract postdoctoral fellows from selected basic science disciplines into the fields of communication sciences. Additional special programs should be devised to encourage established investigators in other basic science disciplines to undertake research on hearing, balance, speech, language, voice, the chemical senses, and other relevant program areas.
- A new small grants program should be introduced for the support of short-term training to learn new technologies. This mechanism could be used by new or established investigators to allow them to learn an area of new technology or a specific research technique available in a few institutions. A three-year trial mechanism should be established to review applications and to award funds within a very short period of time (one to three months). Awards would be small ($1,000 to $3,000) and decided administratively by NIDCD program staff. The resulting productivity of recipients could be monitored with progress reports. Although there would be no pre-award peer review, the National Deafness and Other Communication Disorders Advisory Council would evaluate the effectiveness of the program and determine whether it should be continued beyond the initial three years.
- Some areas of communication and sensory science are relatively new fields, which are in a rapidly growing phase. They require rapid increases in trained investigator personnel resources for undertaking the research recommended in this National Strategic Research Plan.
- The rate at which senior investigators are expected to retire is greater than the current rate of training. The number of basic and clinical investigators in training should therefore be doubled immediately. To take advantage of the many new research areas outlined elsewhere in this report, it may be necessary again to double the total number of investigators in training by the year 2000. An across-the-board doubling or quadrupling in every program area is not proposed. In certain areas, where the research personnel pool is especially limited, a five- to tenfold or even greater increase in numbers may be

advisable. Such fields need special attention to ensure their survival and progress.

- A strong and comprehensive basic and clinical research program in deafness and other communication disorders research must be established at the NIH Clinical Center. There should be ample funding to support the use of these laboratories as a major training ground for the next generation of investigators. Sections should be set up in each of the scientific program areas to focus on the most pressing research needs in each area since the NIDCD is gravely underrepresented at the Clinical Center.
- The lack of epidemiological data must be addressed. Contracts should be offered to alleviate the most pressing needs. For the long term, epidemiological training programs should be established within schools of public health and medical schools having strong programs in epidemiology. These training programs would be specifically designed to train investigators for communication disorders epidemiology. They should be interdisciplinary undertakings involving both epidemiologists and communication scientists.
- The NIDCD should lead the NIH in efforts to recruit and train deaf investigators and clinicians and to assertively pursue the recruitment and research training of individuals with communication disorders. Too often, deafness and communication disorders have been grounds for employment discrimination. The NIDCD has a special responsibility to assure that these citizens are offered equal opportunity to be included in the national biomedical research enterprise.
- The NIDCD should encourage all NIH grantees and personnel who will be interacting with deaf investigators, patients, and the public to learn and use elementary sign language and to become familiar with assistive devices used by the deaf and hearing impaired people. The NIH should be sensitive to the needs of deaf people and individuals with communication disorders in planning and implementing NIH-sponsored advisory meetings and conferences.

NATIONAL INFORMATION CLEARINGHOUSE

The purpose of an information clearinghouse is to collect, categorize, store, and disseminate information that is not otherwise readily accessible to the public and health care professionals. Specific functions of a clearinghouse should be defined by members of the community or communities that it will later serve. In addition to the information collection and dissemination function, clearinghouses are also resources for their communities. Staff members answer telephoned and written inquiries, direct consumers and health care professionals alike to other resources—both private and public, maintain computer data bases, and sponsor workshops and con-

ferences. As part of their information function, clearinghouses identify gaps in information and develop strategies for filling the gaps. Clearinghouses also develop information networks with their primary associations and related groups for meeting, in the most economical manner, the educational and informational needs of their publics.

The Task Force recommends, for meeting the requirements of Public Law 100-553, that the NIDCD establish a planning panel for a clearinghouse (or clearinghouses) at the NIDCD. The Task Force further recommends that the planning panel define the scope of activity and develop guidelines for the NIDCD clearinghouse(s) office, and that the NIDCD establish a Clearinghouse Advisory Panel or panels for the NIDCD clearinghouse office. The planning panel should consider, among other issues, the following questions and concerns:

- In view of the diversity of the disciplines of the NIDCD, should there be several NIDCD information clearinghouses?
- What types of information and knowledge should be processed by the clearinghouse(s), and how does the Clearinghouse Advisory Panel participate in this processing? For instance:
  - scientific data
  - results of epidemiologic studies
  - data from clinical trials
  - patient-related referrals and information
  - interprofessional and intraprofessional interchanges
  - organ donation—for instance, temporal bones and temporal bone bank information
  - sources of research funding—Federal and especially non-Federal
- How should the clearinghouse(s) be coordinated with other data acquisition and dissemination activities at the NIDCD—for instance, the data systems and population-based studies (see above), and the dissemination functions performed by the National Research and Training Centers (see also above)?
- How should the NIDCD clearinghouse(s) be coordinated with other NIH clearinghouses and related activities at the NIH and in the Federal Government, such as MEDLINE? Should there be an interagency mechanism, and should the clearinghouse office have representation in the interagency mechanism?
- How can the clearinghouse(s) assist in the furtherance of research?
- How would the clearinghouse(s) assist in the translation of material for non-English-speaking populations?
- How do the clearinghouse(s) "network" the existing information and knowledge resources?
- How do the clearinghouse(s) obtain, catalog, and disseminate "fugitive" materials?

Members of the planning panel should represent NIDCD specialty areas and also be knowledgeable about information dissemination. The specialty areas that should be represented on the planning panel for the NIDCD clearinghouse(s) are:

- HEARING: Genetics, epidemiology, clinical audiology, otology, industrial

audiology, public school audiology, deaf education

- TASTE/SMELL/TOUCH: Taste, smell, and touch/temperature disorders; psychophysics; genetics; nutrition; and epidemiology
- SPEECH/VOCAL PRODUCTION: Voice, craniofacial anomalies, cleft palate, stuttering, articulation/development, swallowing, laryngology
- VESTIBULAR SYSTEM: Neurology, balance therapy, neuroophthalmology, gerontology
- LANGUAGE: Aphasiology; specialists in developmental language disorders, pediatrics, oralism, total communication, and bilingual approach to language

Members of the planning panel should also represent a mix of organizations and agencies that disseminate information and knowledge about deafness and other communication disorders: Federal and non-Federal agencies, foundations, professional organizations and societies, manufacturers, colleges and universities, hospitals and clinics, local support organizations and chapters, and international organizations and societies. The information officer of the current NINCDS and at least one representative from an existing NIH clearinghouse should serve as advisors to the planning panel. The mix should be drawn from the groups listed in Appendix C.

MULTICULTURAL RESEARCH

The range of scientific and clinical concerns of the NIDCD should encompass the current pluralistic character of society as well as the demographic projections for the nation. Although it is projected that one-third of the nation will be comprised of racial and ethnic minority groups by the turn of the century, only scant reference has been made to racial and ethnic minorities in research on communication disorders. Such research has been limited primarily to phonological and grammatical differences. However, critical areas in need of multicultural research span the full range of topics in communicative disorders.

While minority groups suffer from the same types of communication disorders as nonminority groups, there are pertinent differences in prevalence, etiology, and manifestations of speech, language, and hearing disorders in need of extensive research. Therefore, scientific and clinical emphasis on U.S. black, Hispanic, Asian, and American Indian populations is needed in at least the following areas:

- Basic epidemiologic information is needed on the incidence, prevalence, and risk factors of speech, language, and hearing disorders within black, Hispanic, Asian, and American Indian populations. Most of the available epidemiologic information has been gathered on white populations or, in a few cases, comparatively between

"whites," "blacks," and "others." Such data are not generalizable to specific minority populations.

- Research on fluency within minority populations is sorely needed. For instance, emerging evidence suggests cross-cultural differences in prevalence and in the types of disfluent behaviors. Such findings not only have implications for treatment but can expand the scientific foundations regarding environmental, linguistic, and physiological bases of fluency disorders.
- Voice characteristics and disorders of voice warrant investigation across multicultural populations. Scattered evidence exists regarding anthropomorphic distinctions that underlie cross-racial differences in fundamental frequency. However, the full range of vocal tract resonance characteristics across multicultural populations is in need of study. Similarly, other factors that may underlie cross-racial or cross-cultural differences in vocal pathology require study, such as cultural differences in styles of discourse, occupational differences, dietary differences, perceptions of vocal aesthetics, differences in lifestyle associated with exposure to teratogens, prevalence of upper respiratory disorders, and so on.
- Research is needed on the relationship between communication disorders and various diseases and disorders that have a high prevalence within minority populations. There is only limited research, for instance, on the relationship between communication disorders and such diseases and disorders as sickle cell anemia, excess lead absorption, fetal alcohol syndrome, HIV infection, hypertension, and diabetes, which have a disproportionately high prevalence among minority children and adults.
- Research on normal hearing, hearing loss, deafness, and otological pathologies is needed across multicultural populations. Otitis media occurs most frequently, for instance, among American Indians and least frequently among blacks. Likewise, the black elderly population has a lower prevalence of tinnitus than the white elderly population. Although various explanations have been offered, ranging from cross-racial differences in eustachian tube structure and function to differences in general health status, there is no definitive evidence for such findings.
- Multilingual factors that impact on psychoacoustic perception need research. For instance, recent research suggests that esophageal speakers of tonal languages have poorer intelligibility than esophageal speakers of nontonal languages when judged by native speakers of the respective languages.
- Normative studies focused on the acquisition of phonological and grammatical forms as well as language use and content are needed across multicultural populations. Although psycholinguistic research has moved beyond the analysis of linguistic form, such studies still form the basis of clinical assessment. Most normative data in common use in clinical practice are not generalizable to nonmainstream populations.

- Similar to the need for culturally appropriate normative data is the need for nondiscriminatory assessment procedures. Alternatives to the use of standardized procedures must be studied for use in the clinical assessment of individuals from multicultural populations.
- Factors that impede efficacy of treatment in the clinical management of multicultural populations require study. Cross-cultural differences in cognitive/learning styles, interpersonal and ecological factors that affect the clinical encounter, cultural systems of belief that may be in conflict with approaches to treatment, and other related factors need research.

There must be scrutiny of the types of research paradigms that are proposed for studying multicultural populations. There has almost been a preoccupation with the comparative paradigm for studying minority groups. Alternate research designs such as retrospective designs and path analysis offer greater ethnographic validity and applicability than comparative models.

In addition to these concerns, some members of the Task Force discussed ethical aspects of the psychosocial effects of communication impairments and their remediation, especially from the perspective of impairment as a culturally defining condition. It was suggested that research on these effects is needed. They include problems of special education placement versus "mainstreaming," factors contributing to stigmatization, and fair and equitable distribution of remediation services and communication aids and devices across multicultural and socioeconomic groups.

Appendix A.

Public Law 100 - 553

Public Law 100-553
100th Congress

An Act

To amend the Public Health Service Act to establish within the National Institutes of Health a National Institute on Deafness and Other Communication Disorders.

Oct. 28, 1988
[S. 1727]

Be it enacted by the Senate and House of Representatives of the United States of America in Congress assembled,

National Deafness and Other Communication Disorders Act of 1988.
42 USC 201 note.

SECTION 1. SHORT TITLE.

This Act shall be cited as the "National Deafness and Other Communication Disorders Act of 1988".

SEC. 2. ESTABLISHMENT AND TRANSFER OF FUNCTIONS.

Title IV of the Public Health Service Act (42 U.S.C. 281 et seq.) is amended—

(1) in section 401(b)(1)—

(A) by striking "and Communicative" in subparagraph (J); and

(B) by adding at the end the following new subparagraph:

"(M) The National Institute on Deafness and Other Communication Disorders.";

(2) in the heading for subpart 10 of part C, by striking "and Communicative";

(3) in section 457—

(A) by striking "and Communicative"; and

(B) by striking "disorder, stroke," and all that follows and inserting "and disorder and stroke."; and

(4) in Part C, by adding at the end the following new subpart:

"Subpart 13—National Institute on Deafness and Other Communication Disorders

Education.
Research and development.
Public information.

"PURPOSE OF THE INSTITUTE

"SEC. 464. The general purpose of the National Institute on Deafness and Other Communication Disorders (hereafter referred to in this subpart as the 'Institute') is the conduct and support of research and training, the dissemination of health information, and other programs with respect to disorders of hearing and other communication processes, including diseases affecting hearing, balance, voice, speech, language, taste, and smell.

42 USC 285m.

"NATIONAL DEAFNESS AND OTHER COMMUNICATION DISORDERS PROGRAM

"SEC. 464A. (a) The Director of the Institute, with the advice of the Institute's advisory council, shall establish a National Deafness and Other Communication Disorders Program (hereafter in this section referred to as the 'Program'). The Director or the Institute shall, with respect to the Program, prepare and transmit to the Director of NIH a plan to initiate, expand, intensify and coordinate activities of the Institute respecting disorders of hearing (including tinnitus) and

42 USC 285m-1.

29-139 (553)

other communication processes, including diseases affecting hearing, balance, voice, speech, language, taste, and smell. The plan shall include such comments and recommendations as the Director of the Institute determines appropriate. The Director of the Institute shall periodically review and revise the plan and shall transmit any revisions of the plan to the Director of NIH.

"(b) Activities under the Program shall include—

"(1) investigation into the etiology, pathology, detection, treatment, and prevention of all forms of disorders of hearing and other communication processes, primarily through the support of basic research in such areas as anatomy, audiology, biochemistry, bioengineering, epidemiology, genetics, immunology, microbiology, molecular biology, the neurosciences, otolaryngology, psychology, pharmacology, physiology, speech and language pathology, and any other scientific disciplines that can contribute important knowledge to the understanding and elimination of disorders of hearing and other communication processes;

"(2) research into the evaluation of techniques (including surgical, medical, and behavioral approaches) and devices (including hearing aids, implanted auditory and nonauditory prosthetic devices and other communication aids) used in diagnosis, treatment, rehabilitation, and prevention of disorders of hearing and other communication processes;

"(3) research into prevention, and early detection and diagnosis, of hearing loss and speech and language disturbances (including stuttering) and research into preventing the effects of such disorders on learning and learning disabilities with extension of programs for appropriate referral and rehabilitation;

"(4) research into the detection, treatment, and prevention of disorders of hearing and other communication processes in the growing elderly population with extension of rehabilitative programs to ensure continued effective communication skills in such population;

"(5) research to expand knowledge of the effects of environmental agents that influence hearing or other communication processes; and

"(6) developing and facilitating intramural programs on clinical and fundamental aspects of disorders of hearing and all other communication processes.

"DATA SYSTEM AND INFORMATION CLEARINGHOUSE

42 USC 285m-2.

"SEC. 464B. (a) The Director of the Institute shall establish a National Deafness and Other Communication Disorders Data System for the collection, storage, analysis, retrieval, and dissemination of data derived from patient populations with disorders of hearing or other communication processes, including where possible, data involving general populations for the purpose of identifying individuals at risk of developing such disorders.

"(b) The Director of the Institute shall establish a National Deafness and Other Communication Disorders Information Clearinghouse to facilitate and enhance, through the effective dissemination of information, knowledge and understanding of disorders of hearing and other communication processes by health professionals, patients, industry, and the public.

"MULTIPURPOSE DEAFNESS AND OTHER COMMUNICATION DISORDERS CENTER

"SEC. 464C. (a) The Director of the Institute shall, after consultation with the advisory council for the Institute, provide for the development, modernization, and operation (including care required for research) of new and existing centers for studies of disorders of hearing and other communication processes. For purposes of this section, the term 'modernization' means the alteration, remodeling, improvement, expansion, and repair of existing buildings and the provision of equipment for such buildings to the extent necessary to make them suitable for use as centers described in the preceding sentence. 42 USC 285m-3.

"(b) Each center assisted under this section shall—

"(1) use the facilities of a single institution or a consortium of cooperating institutions; and

"(2) meet such qualifications as may be prescribed by the Secretary.

"(c) Each center assisted under this section shall, at least, conduct—

"(1) basic and clinical research into the cause diagnosis, early detection, prevention, control and treatment of disorders of hearing and other communication processes and complications resulting from such disorders, including research into rehabilitative aids, implantable biomaterials, auditory speech processors, speech production devices, and other otolaryngologic procedures;

"(2) training programs for physicians, scientists, and other health and allied health professionals; Health care professionals.

"(3) information and continuing education programs for physicians and other health and allied health professionals who will provide care for patients with disorders of hearing or other communication processes; and

"(4) programs for the dissemination to the general public of information—

"(A) on the importance of early detection of disorders of hearing and other communication processes, of seeking prompt treatment, rehabilitation, and of following an appropriate regimen; and

"(B) on the importance of avoiding exposure to noise and other environmental toxic agents that may affect disorders of hearing or other communication processes.

"(d) A center may use funds provided under subsection (a) to provide stipends for health professionals enrolled in training programs described in subsection (c)(2).

"(e) Each center assisted under this section may conduct programs—

"(1) to establish the effectiveness of new and improved methods of detection, referral, and diagnosis of individuals at risk of developing disorders of hearing or other communication processes; and

"(2) to disseminate the results of research, screening, and other activities, and develop means of standardizing patient data and recordkeeping.

"(f) The Director of the Institute shall, to the extent practicable, provide for an equitable geographical distribution of centers assisted under this section. The Director shall give appropriate consideration Aged persons. Children and youth.

to the need for centers especially suited to meeting the needs of the elderly, and of children (particularly with respect to their education and training), affected by disorders of hearing or other communication processes.

"(g) Support of a center under this section may be for a period not to exceed seven years. Such period may be extended by the Director of the Institute for one or more additional periods of not more than five years if the operations of such center have been reviewed by an appropriate technical and scientific peer review group established by the Director, with the advice of the Institute's advisory council, if such group has recommended to the Director that such period should be extended.

"NATIONAL INSTITUTE ON DEAFNESS AND OTHER COMMUNICATION DISORDERS ADVISORY BOARD

42 USC 285m-4.

"SEC. 464D. (a) The Secretary shall establish in the Institute the National Deafness and Other Communication Disorders Advisory Board (hereafter in this section referred to as the 'Advisory Board').

"(b) The Advisory Board shall be composed of eighteen appointed members and nonvoting ex officio members as follows:

"(1) The Secretary shall appoint—

"(A) twelve members from individuals who are scientists, physicians, and other health and rehabilitation professionals, who are not officers or employees of the United States, and who represent the specialties and disciplines relevant to deafness and other communication disorders, including not less than two persons with a communication disorder; and

"(B) six members from the general public who are knowledgeable with respect to such disorders, including not less than one person with a communication disorder and not less than one person who is a parent of an individual with such a disorder.

Of the appointed members, not less than five shall by virtue of training or experience be knowledgeable in diagnoses and rehabilitation of communication disorders, education of the hearing, speech, or language impaired, public health, public information, community program development, occupational hazards to communications senses, or the aging process.

"(2) The following shall be ex officio members of each Advisory Board:

"(A) The Assistant Secretary for Health, the Director of NIH, the Director of the National Institute on Deafness and Other Communication Disorders, the Director of the Centers for Disease Control, the Chief Medical Director of the Veterans' Administration, and the Assistant Secretary of Defense for Health Affairs (or the designees of such officers).

"(B) Such other officers and employees of the United States as the Secretary determines necessary for the Advisory Board to carry out its functions.

"(c) Members of an Advisory Board who are officers or employees of the Federal Government shall serve as members of the Advisory Board without compensation in addition to that received in their regular public employment. Other members of the Board shall receive compensation at rates not to exceed the daily equivalent of

the annual rate in effect for grade GS-18 of the General Schedule for each day (including traveltime) they are engaged in the performance of their duties as members of the Board.

"(d) The term of office of an appointed member of the Advisory Board is four years, except that no term of office may extend beyond the expiration of the Advisory Board. Any member appointed to fill a vacancy for an unexpired term shall be appointed for the remainder of such term. A member may serve after the expiration of the member's term until a successor has taken office. If a vacancy occurs in the Advisory Board, the Secretary shall make an appointment to fill the vacancy not later than 90 days from the date the vacancy occurred.

"(e) The members of the Advisory Board shall select a chairman from among the appointed members.

"(f) The Secretary shall, after consultation with and consideration of the recommendations of the Advisory Board, provide the Advisory Board with an executive director and one other professional staff member. In addition, the Secretary shall, after consultation with and consideration of the recommendations of the Advisory Board, provide the Advisory Board with such additional professional staff members, such clerical staff members, such services of consultants, such information, and (through contracts or other arrangements) such administrative support services and facilities, as the Secretary determines are necessary for the Advisory Board to carry out its functions.

"(g) The Advisory Board shall meet at the call of the chairman or upon request of the Director of the Institute, but not less often than four times a year.

"(h) The Advisory Board shall—

"(1) review and evaluate the implementation of the plan prepared under section 464A(a) and periodically update the plan to ensure its continuing relevance;

"(2) for the purpose of assuring the most effective use and organization of resources respecting deafness and other communication disorders, advise and make recommendations to the Congress, the Secretary, the Director of NIH, the Director of the Institute, and the heads of other appropriate Federal agencies for the implementation and revision of such plan; and

"(3) maintain liaison with other advisory bodies related to Federal agencies involved in the implementation of such plan and with key non-Federal entities involved in activities affecting the control of such disorders.

"(i) In carrying out its functions, the Advisory Board may establish subcommittees, convene workshops and conferences, and collect data. Such subcommittees may be composed of Advisory Board members and nonmember consultants with expertise in the particular area addressed by such subcommittees. The subcommittees may hold such meetings as are necessary to enable them to carry out their activities.

"(j) The Advisory Board shall prepare an annual report for the Secretary which— Reports.

"(1) describes the Advisory Board's activities in the fiscal year for which the report is made;

"(2) describes and evaluates the progress made in such fiscal year in research, treatment, education, and training with respect to the deafness and other communication disorders;

"(3) summarizes and analyzes expenditures made by the Federal Government for activities respecting such disorders in such fiscal year; and

"(4) contains the Advisory Board's recommendations (if any) for changes in the plan prepared under section 464A(a).

"(k) The National Deafness and Other Communication Disorders Advisory Board shall be established not later than 90 days after the date of the enactment of the National Institute on Deafness and Other Communication Disorders Act.

"INTERAGENCY COORDINATING COMMITTEE

42 USC 285m-5.

"SEC. 464E. (a) The Secretary may establish a committee to be known as the Deafness and Other Communication Disorders Interagency Coordinating Committee (hereafter in this section referred to as the 'Coordinating Committee').

"(b) The Coordinating Committee shall, with respect to deafness and other communication disorders—

"(1) provide for the coordination of the activities of the national research institutes; and

"(2) coordinate the aspects of all Federal health programs and activities relating to deafness and other communication disorders in order to assure the adequacy and technical soundness of such programs and activities and in order to provide for the full communication and exchange of information necessary to maintain adequate coordination of such programs and activities.

"(c) The Coordinating Committee shall be composed of the directors of each of the national research institutes and divisions involved in research with respect to deafness and other communication disorders and representatives of all other Federal departments and agencies whose programs involve health functions or responsibilities relevant to deafness and other communication disorders.

"(d) The Committee shall be chaired by the Director of NIH (or the designee of the Director). The Committee shall meet at the call of the chair, but not less often than four times a year.

Reports.

"(e) Not later than 120 days after the end of each fiscal year, the Committee shall prepare and transmit to the Secretary, the Director of NIH, the Director of the Institute, and the advisory council for the Institute a report detailing the activities of the Committee in such fiscal year in carrying out subsection (b).

"LIMITATION ON ADMINISTRATIVE EXPENSES

42 USC 285m-6.

"SEC. 464F. With respect to amounts appropriated for a fiscal year for the National Institutes of Health, the limitation established in section 408(b)(1) on the expenditure of such amounts for administrative expenses shall apply to administrative expenses of the National Institute on Deafness and Other Communication Disorders.".

42 USC 285m note.

SEC. 3. TRANSITIONAL AND SAVINGS PROVISIONS.

Gifts and property. Contracts. Records.

(a) TRANSFER OF PERSONNEL, ASSETS, AND LIABILITIES.—Personnel employed by the National Institutes of Health in connection with the functions vested under section 2 in the Director of the National Institute on Deafness and Other Communication Disorders, and assets, property, contracts, liabilities, records, unexpended balances of appropriations, authorizations, allocations, and other funds of the National Institutes of Health, arising from or employed, held, used,

available to, or to be made available, in connection with such functions shall be transferred to the Director for appropriate allocation. Unexpended funds transferred under this subsection shall be used only for the purposes for which the funds were originally authorized and appropriated.

(b) SAVINGS PROVISIONS.—With respect to functions vested under section 1 in the Director of the National Institute on Deafness and Other Communication Disorders, all orders, rules, regulations, grants, contracts, certificates, licenses, privileges, and other determinations, actions, or official documents, that have been issued, made, granted, or allowed to become effective, and that are effective on the date of the enactment of this Act, shall continue in effect according to their terms unless changed pursuant to law. Grants. Contracts.

Approved October 28, 1988.

LEGISLATIVE HISTORY—S. 1727 (H.R. 3361):

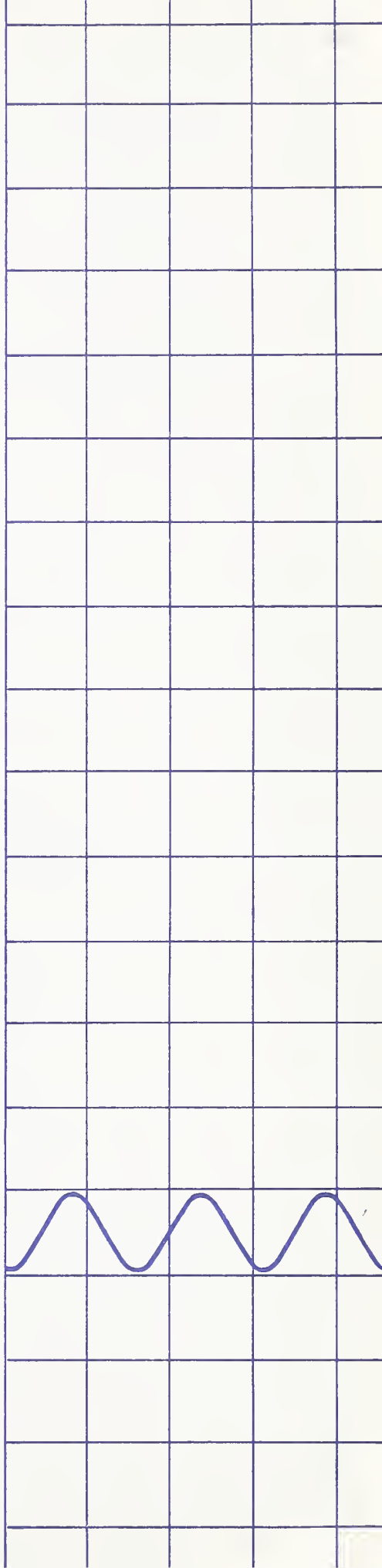
HOUSE REPORTS: No. 100-761 accompanying H.R. 3361 (Comm. on Energy and Commerce).

CONGRESSIONAL RECORD, Vol. 134 (1988):
Oct. 7, considered and passed Senate.
Oct. 13, considered and passed House.

○

Appendix B.

Task Force on the National Strategic Research Plan



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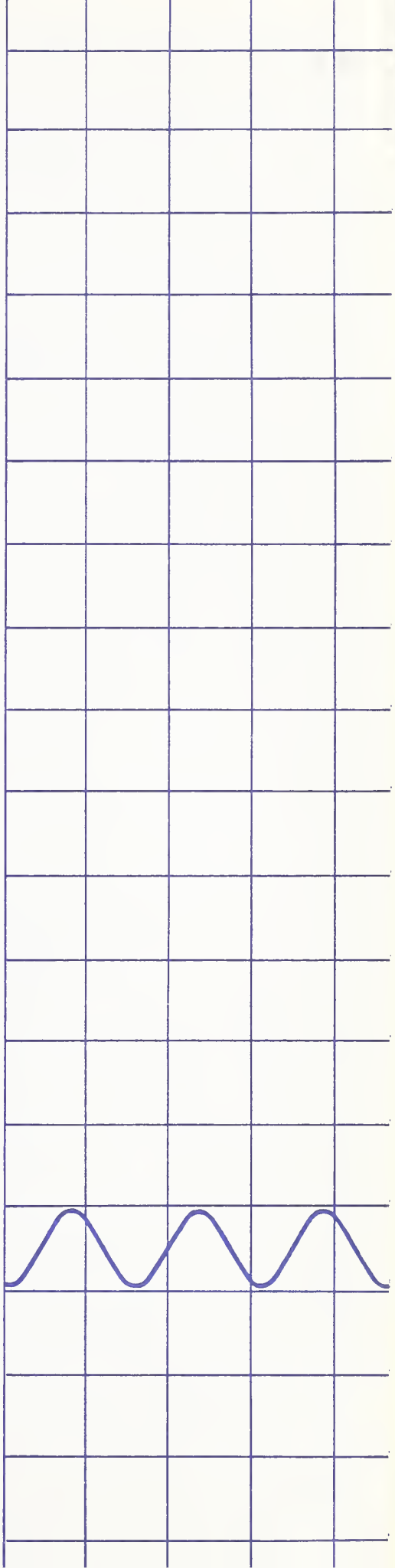
Appendix C.

Reviewing Organizations



REVIEWING ORGANIZATIONS

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Minneapolis, MN

Alexander Graham Bell Association
Washington, DC

American Academy of Otolaryngology
Washington, DC

American Association of the Deaf-Blind
Silver Spring, MD

American Auditory Society
Dallas, TX

American Cleft Palate Association—National Office
Pittsburgh, PA

American Deafness and Rehabilitation Association
Washington, DC

American Hearing Research Foundation
Chicago, IL

American Society for Deaf Children
Silver Spring, MD

American Speech-Language-Hearing Association
Rockville, MD

American Tinnitus Association
Portland, OR

Association for Research in Otolaryngology
Boston, MA

Association for Chemoreception Sciences
Philadelphia, PA

Better Hearing Institute
Washington, DC

Central Institute for the Deaf
St. Louis, MO

Committee on Hearing Bioacoustics and Biomechanics—National Academy of Sciences
Washington, DC

Convention of American Instructors of the Deaf
Columbia, MD

Deafness Research Foundation
New York, NY

Duncan & Associates
Washington, DC

Gallaudet University—Conference of Educational Administrators Serving the Deaf/Pre-College Programs
Washington, DC

Hearing Industries Association
Washington, DC

International Society for Alternative and Augmentative Communication
Toronto, Canada

Meniere's Network
Nashville, TN

National Association of the Deaf
Silver Spring, MD

National Fraternal Society of the Deaf
Seabrook, MD

National Captioning Institute
Falls Church, VA

National Hearing Aid Society
Lavonia, MI

National Society for Children and Adults with Autism
Washington, DC

National Aphasia Association
New York, NY

National Foundation for Children's Hearing Education and Research
Yonkers, NY

National Technical Institute for the Deaf
Rochester, NY

Self Help for Hard of Hearing People
Bethesda, MD

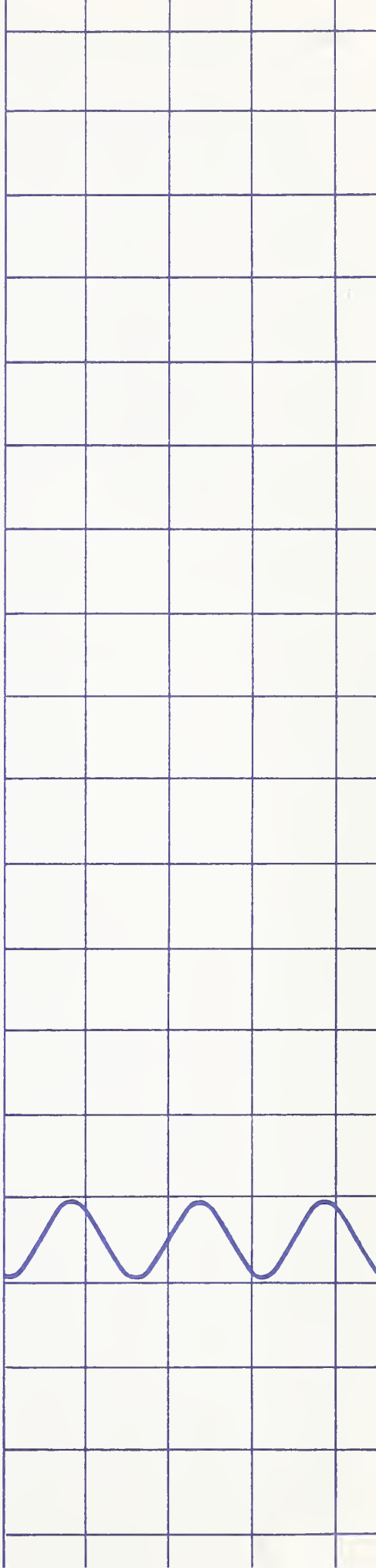
Telecommunications for the Deaf, Inc.
Silver Spring, MD

Tourette's Syndrome Association
Bayside, NY

Voice Foundation
New York, NY

Appendix D.

Comments by Reviewing Organizations



COMMENTS BY REVIEWING ORGANIZATIONS

A prepublication draft of this report was sent to the organizations listed in Appendix C for suggestions and comments. Their responses, which are addressed to that draft, are reprinted here as a matter of information. In some instances, it was possible to edit the text to meet their concerns. The Institute and its advisors will consider these comments and incorporate them into the final research plan.

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21 March 1989

Sean Donohue
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Dear Mr. Donohue:

Thank you for the invitation to comment on the Draft Report on the National Strategic Research Plan for NIDCD. It is indeed an excellent document, and should serve well its intended purpose.

I have found only a few minor flaws in the section on Deafness and Hearing Disorders, and but one major one. The major problem occurs on page 2-20, in the first section "Susceptibility to Environmentally Caused Deafness". The first sentence indicates that deterioration of hearing with time is a combination of the effects of industrial noise; of the noises of everyday living; of diseases, drugs, chemicals and trauma; and of the aging process per se. Notably missing from this list is the degeneration of hearing with age at a rate far in excess of what can be ascribed to the aging process, in the absence of any known high-intensity acoustic stimulation, diseases, drugs, chemicals or trauma; in the past this has generally been called "progressive sensorineural loss of unknown etiology", although in some cases an hereditary factor was strongly indicated by occurrence in many members of the same family.

However, the problem is not solved by inventing a new term, "mixacusis", to define that type of hearing loss, especially a term that has, as far as I have been able to determine, no etymological relation to the phenomenon concerned. Perhaps "heredicusis" or "genetocusis" might serve; however, the present document is hardly the place to introduce it. And as it is used in sentence 2, it isn't even what it is intended to be; as it stands, "mixacusis" is "one's genetic predisposition", rather than "hearing loss due to genetic predisposition". Furthermore, it is the genetic predisposition, not the hearing loss it would produce by itself, that does "interact" with the preceding factors. The problem evaporates if the term "mixacusis" is simply deleted. I entreat you to do so.

Minor: in that same paragraph, third line from end, it should read "..... susceptibility to hearing loss...", not "for".

I hope I have been persuasive.

Sincerely,

W. Dixon Ward
President, ASA

Alexander Graham Bell
Association for the Deaf, Inc.

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Established in 1890 to encourage the use of residual hearing and the teaching of speech and speechreading.

Office of the Executive Director

April 18, 1989

Mr. Sean Donohue
National Institutes of Health
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Dear Mr. Donohue:

On behalf of the Alexander Graham Bell Association for the Deaf, we are pleased to have the opportunity to review the draft report of the Task Force on the National Strategic Research Plan for the National Institute on Deafness and Other Communicative Disorders. It is clear from the magnitude of the document that the Task Force members worked hard to develop this report and the resulting recommendations for research in the topic areas of deafness and hearing disorders; balance and the vestibular system; voice and voice disorders; speech and speech disorders; language and language impairments; and taste, smell, and touch.

We shall focus our remarks on the Chapters and/or appendices which are particularly relevant to the concerns of the Bell Association. Our membership is composed of professionals involved in working with hearing-impaired individuals; parents of hearing impaired youngsters; and hearing-impaired (deaf and hard of hearing) adults. Our comments have been formulated after review of the draft of the research plan; review of the titles of the on-going research projects currently underway at NIDCD; discussion with our Executive Committee and with some of our membership directly involved in research and discussion with the membership of the Council of Organizational Representatives (COR).

We recognize that the medical model of the draft National Strategic Research Plan is consistent with existing NIH Institutes. However, as our colleague Rocky Stone, Executive Director of Self Help for Hard of Hearing People, so eloquently states on page 3 of his response to the draft:

"The Surgeon General of the United States has stated that 'Mending people, curing them, will no longer be enough. That is only part of the total health care that most people require.' Unless this thinking is incorporated into NIDCD's philosophy at an early stage, the Institute's research will have less impact than its potential warrants. In the hearing health field NIDCD could forge the first usable model to provide a basis for the understanding, treatment and rehabilitation of hearing impaired persons, the goal which the American Academy of Otolaryngology-Head and Neck Surgery (AAO) projects as its mandate."

We would support this innovative, comprehensive approach to the mission of the National Institute on Deafness and Other Communicative Disorders.

Since our Association is strongly committed to every hearing-impaired child having the opportunity to develop spoken language, we are pleased to see that identification and description of patterns of language acquisition of children exposed primarily to oral language is cited as an area for continued research. Further, we would welcome additional research directed at a better understanding of the typical patterns of spoken language development of deaf children of both hearing and deaf parents. We are particularly interested in increasing knowledge of those factors (physiological, psychological, environmental, etc.) which lead to successful acquisition of spoken language in both prelingually and post lingually hearing-impaired children.

The Bell Association is extremely concerned about the issue of literacy and supports increased research in this area. It would seem appropriate to continue to evaluate the training and stimulation programs that result in so many oral hearing-impaired youngsters abilities to perform at grade level and above. This should be a specific area for significant research within the strategic plan.

The Bell Association's interests in hearing-impairment span the lifetime of a hearing-impaired individual as well as issues related to both hard of hearing and deaf persons. Three areas of long standing great concern to our Association are issues of prevention, early identification, and intervention. Thus, we view genetic studies, assessment of ototoxic impact of viral diseases, blood incompatibility studies and evaluation of all other factors that potentially place a newborn at high risk for hearing loss as most important research areas for NIDCD. Obviously, several of the research areas apply to older hearing-impaired persons as well. In addition, we are supportive of further study of cause and treatment of presbycusis, tinnitus, Menieres disease, noise-induced hearing loss and sudden hearing loss of all types.

Investigations related to hearing impairment and its impact on all aspects of both the hearing-impaired individual's life and

the lives of family members are of concern to our Association. Thus the analysis of the relationships of stress, depression, substance abuse, employment and family problems to hearing-impairment should be a part of the strategic research plan.

Technological advances hold great promise for helping hearing-impaired individuals of all ages. We support research on cochlear implants, virbo-tactile devices, and assistive listening devices of all types, INCLUDING HEARING AIDS

Finally, in establishing clearinghouses we urge "networking" with existing information and knowledge resources as stated on page 12-2. The Alexander Graham Bell Association for the Deaf was founded 99 years ago for the "diffusion of knowledge" about hearing loss. Thus it seems appropriate to suggest that we and other longstanding consumer organizations within the COR structure should be participants on the planning panel for the clearinghouses so that all existing resources may be coordinated.

Thank you for the opportunity to review the draft National Strategic Research Plan. We look forward to the time when the Institute will have completed the necessary stages of formulation and entered the period focused on investigating and answering many of the questions posed in the Strategic Research Plan.

Sincerely,

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Dear Mr. Donohue:

On behalf of the American Speech-Language-Hearing Association (ASHA), the national scientific and professional association representing 57,000 speech-language pathologists and audiologists, I would like to thank you for the opportunity to comment on the draft document "The National Strategic Research Plan for the National Institute on Deafness and Other Communication Disorders."

Before providing any specific comments I would like to commend Dr. Jay Moskowitz, Acting Director of the Institute, and the task force members for their dedication and diligence in producing such a fine document, especially under such time constraints. Both the process and the product augur a fine beginning for the National Institute on Deafness and Other Communication Disorders (NIDCD). This document will provide strong guidance for the implementation of a research plan for the Institute.

An area addressed in the document and one of particular concern to ASHA is that of training. With the small cadre of researchers in the speech-language-hearing profession there is a need both to nurture and support current researchers and also to encourage new investigators to devote their careers to research. Training is needed particularly at the predoctoral level to provide a firm foundation from which accomplished researchers can emanate. The Association is acutely aware of the limitations on funds for training with the National Institutes of Health (NIH). However, for this new Institute the training function will be one that is especially needed to foster current and to found future research. The National Research and Training Centers offer one vehicle for providing training; however, clearly there needs to be additional monies allocated to training outside these centers. Additional funds are needed for training beyond those requested in the 1990 FY budget. The Association would be interested in collaborating with NIDCD on endeavors examining training models to maximize their effectiveness.

The National Research and Training Centers as described in the document are different from past NIH Centers of Excellence. Their description in PL 100-553 specifies numerous roles for a center, with the effect that excellent candidates as centers might be excluded because of failure to meet all the roles. ASHA would recommend that the interpretation of the law be very liberal in this regard and that centers not be required to fulfill so many roles as to dilute their effectiveness. With a new institute, it would seem prudent to establish such centers gradually. When funding is at a sufficient level for individual investigator awards, such as R01s, then centers could begin to be phased in. The Association is concerned that monies not be diverted from R01 grants to centers, such that the funding level of R01 grants would be compromised.

The Association does have some experience with clearinghouse functions through our ACTIONLINE and Public Information Department. We would be happy to offer our experience in this regard should it be helpful.

Because all scientific panels at the task force meeting did not have an opportunity to develop a detailed research program and because that was not part of the charge to each scientific panel, it would seem that having the "Comprehensive Program of Research on Deafness and Hearing Disorders" available on request rather than as a part of the document would balance the report across all the scientific disciplines.

I am aware that Dr. Cynthia Shewan, Director of our Research Division, has sent you some editorial changes under separate cover. Therefore, my comments have been limited to the larger issues.

Again, may I congratulate you on production of a comprehensive and excellent document of which NIDCD can be proud. ASHA has enjoyed the privilege of participating in this historic occasion and reaffirms its commitment to assisting the new Institute in any way that it can.

Sincerely,

Gilbert R. Herer, Ph.D.

American Tinnitus Association

P.O. Box 5 • Portland, Oregon 97207 • (503) 248-9985

April 6, 1989

Mr. Sean Donohue
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Bethesda, MD 20892

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Dear Mr. Donohue;

We appreciate the opportunity to comment on the draft of the National Strategic Research Plan for the National Institute on Deafness and Other Communication Disorders. We have limited our comments to issues relating to tinnitus.

First of all I want to call to your attention paragraph 2, page 1-1, which states, "Investigation into the etiology, pathology, detection, treatment and prevention of all forms of disorders of hearing and other communication processes through the support of basic research in such areas as.....". Strictly speaking, it would be difficult if not impossible to investigate treatment and prevention through 'basic research'. Please consider ammending this statement to say "basic and clinical research".

Tinnitus is mentioned on page 1-3, in the paragraph describing chapter 3 . Since tinnitus is not discussed in chapter 3, but earlier, in chapter 2, I believe it would be more appropriate to include tinnitus in the description of chapter 2.

The next mention of tinnitus is on page 2-16 where statistics about tinnitus prevalence have been used. The numbers referred to conflict with published studies from the Hearing Status and Ear Examination report published in 1968 which stated that 26.8% of adults suffer from mild tinnitus and 5.6% suffer from severe tinnitus, and with later reports from the National Health Interview series indicating smaller percentages of sufferers. The NHIS studies relied on self-report, or second party census reports from other family members and would, therefore, be expected to produce different numbers than those from an actual examination. Recent studies from other developed countries indicate numbers closer to the HSEE percentages. For example a study done within the last 10 years in Great Britain and reported in various publications, indicates about 8% of the population suffers from moderate to severe tinnitus, and figures reported from a Swedish study in 1987 indicate 14.2% of the population reporting tinnitus "often or always", and 2.4% saying "tinnitus plagues me all day". The National Health Interview Survey report (1986) provides a tinnitus estimate of 6,315,000, a figure which probably represents severe tinnitus because of the sampling procedures referred to previously. It is unlikely that the prevalence of tinnitus would have lessened in the last 20 years considering the abundance of noise in modern society. This discussion points out the difficulties that we all experience in trying to estimate

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tinnitus prevalence from present sources. Please provide documentation for the figures that are ultimately used in the plan.

We agree that the types and degrees of disability caused by tinnitus need to be characterized and measured in population surveys. It will be extremely important to ask the right questions when these studies are developed. (ATA will be happy to assist researchers with tinnitus questionnaire development, or with information from a survey completed by over 13,000 of our members.) However, many of these data are presently available from a large clinic population. The source of the data is the TDR (Tinnitus Data Registry) of the tinnitus clinic at the Oregon Hearing Research Center. This registry was supported initially by ATA and subsequently by NIH and later grants from the Department of Education. It would also be appropriate for you to list this source, and the prevalence studies as well, on page 10-3. (citations enclosed)

On page 2-16, in the paragraph which begin "Aspirin toxicity... and goes on to state that animal mocels can be used to test tinnitus masking, some conceptual problems arise because current animal models for tinnitus utilize salicylates to induce tinnitus. It has been clinically shown that aspirin induced tinnitus is extremely susceptible to external masking, almost always producing residual inhibition. This would make it troublesome to try to use that particular type of tinnitus for assessing clinical treatment. Furthermore, aspirin toxicity is only one facet of a large issue. What about other drugs that cause tinnitus? What about drugs that change or relieve tinnitus?

Some reliable measures of tinnitus severity are presently in use. (see TDR above) We agree that further measures should be developed in order to measure outcomes. For example, tinnitus appears to mimic other chronic ailments in producing identifiable patterns in personality inventories such as the MMPI. We would like to see further studies utilizing measures of this type to help identify patients in need of special treatment or counseling.

Another important issue that is not addressed in the plan is the relationship between dental conditions and tinnitus. There is ample evidence to suggest such a relationship, particularly between TMJ dysfunction and tinnitus. It is also important to be cognizant of special problems that confront the approximately 10% of tinnitus sufferers who exhibit no measurable hearing loss.

Animal models for tinnitus are desirable and surely will be useful. NIH is presently supporting work on a behavioral animal model for tinnitus. We commend this and hope that studies will be encouraged for a physiological animal model as well. A method is needed for the direct measurement of neurological changes indicating tinnitus. Work along this line has taken place in West Germany and in the U. S. utilizing topographic brain mapping to help elucidate mechanisms of tinnitus.

Appendix A, page 12, lists a research plan for tinnitus. In accordance with the comments above, the priorities implied in the listing may need rearrangement and augmentation.

While recognizing the intrinsic value of basic research I would be remiss in my duty to the 175,000 ATA members if I did not urge you to consider the importance of clinical research to provide more immediate answers to the problem of tinnitus. Research

is needed to:

- Evaluate and refine therapeutic processes
- Search for and identify underlying mechanisms of tinnitus
- Develop both behavioral and physiological animal models for tinnitus
- Better understand the relationship between various drugs and tinnitus
- Better understand the relationship between dental anomalies and tinnitus
- Develop effective, standardized clinical measures of tinnitus sensation
- Develop or refine present measures of tinnitus severity and psychological consequences.
- Initiate prevalence studies for tinnitus in the general population utilizing information already available from existing tinnitus surveys conducted on segments of the tinnitus population.
- Initiate public awareness programs to control noise exposure, particularly directed at the school-age population. (More than half of tinnitus sufferers have significant noise exposure histories.)

Please feel free to call on us at any time for information or assistance. We are vitally interested in the new institute and its objectives. The opportunity is here in this new institute to accelerate efforts on behalf of the hearing impaired, the deaf, and those with other communication disorders. We are counting on you to do the best possible job and we want to help.

Sincerely,

Gloria E. Reich

Gloria E. Reich, Ph.D.
Executive Director

enc. (citations)

References

Axelsson, A., Prevalence of tinnitus, Unpublished manuscript, University of Gothenberg, Sweden, 1987.

Coles, R. R. A. Epidemiology of tinnitus. in Evered, D. (ed.) Tinnitus, Ciba Foundation Symposium 85. Pitman, London, 1981.

Coles, R. R. A. Epidemiology of tinnitus: (1) Prevalence, in A. Shulman and J. Ballantyne, (eds.) Proceedings of the II International Tinnitus Seminar New York, 10-11 June 1983. Supp.9, Jrl Laryn & Oto, 1984.

CENTRAL INSTITUTE FOR THE DEAF
AT WASHINGTON UNIVERSITY MEDICAL CENTER

APR 24 REC'D

April 20, 1989

Jay Moskowitz, Ph.D., Acting Director
National Institute for Deafness
and Other Communicative Disorders
The Shannon Building
Bethesda, MD 20205

Dear Dr. Moskowitz:

Enclosed please find comments prepared by me and two colleagues at Central Institute for the Deaf on the National Strategic Plan of the NIDCD. We feel that the task force has done a good job in pulling together diverse interests and needs within the community served by the NIDCD.

While our comments are brief, each of us has read the report in detail and our notes reflect our common thinking on the National Strategic Plan. We look forward with great anticipation towards continuing our strong cooperation with the NIH as the plan is implemented.

Sincerely yours,

Richard G. Stoker, Ph.D.
Director

Jean S. Moog
School Principal

Ann E. Geers
Director of Clinical Services

307

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Comments on the National Strategic Plan for the National Institute on Deafness and Other Communication Disorders

With regard to the handicapping condition of early childhood deafness, research goals are appropriately targeted toward the development and evaluation of new technology designed to reduce the devastating impact of hearing-impairment. However, the task force should emphasize the importance of going beyond evaluation of improvements in speech perception achieved with new devices to consider broader effects of these devices on developing the ability of deaf individuals to communicate through speech with society at large and to read and write English.

We would recommend a greater emphasis on analysis of overall speech intelligibility, with studies directed toward developing and evaluating ways to capitalize on the new technology for improving speech. Additional studies might be directed toward determining the kinds of devices that might better help deaf individuals monitor their own speech productions. All of these would contribute to improved production of conversational speech of deaf persons.

Regarding educational methodology, topics for research have been thoughtfully outlined. However, we would recommend that rather than focusing on which is best (oral, TC, cued speech, ASL), research should be focused on the advantages or disadvantages of a particular methodology for some particular purpose; e.g. developing spoken language, developing communication, developing ability to read and write English, making money as an adult, developing friends, etc. It is likely that no one methodology is appropriate for all children, and that no one methodology is superior for all of these purposes.

We hope these comments are helpful. We believe that the National Strategic Plan provides a framework for developing important research in deafness.

THE COUNCIL OF
ORGANIZATIONAL REPRESENTATIVES ★

April 18, 1989

Mr. Sean Donohue
National Institutes of Health
Shannon Building, Room 218
9000 Rockville Pike
Bethesda, MD 20892

Dear Mr. Donohue:

This letter serves as a brief summary statement by the Council of Organizational Representatives (COR) to the National Strategic Research Plan prepared by the NIDCD's Task Force.

COR, by way of brief comment, is a coalition of organizations on public policy and other issues related to improving the quality of life for deaf and hard of hearing people. Our membership includes the following eleven organizations:

Alexander Graham Bell Association for the Deaf
American Academy of Otolaryngology-Head and Neck Surgery, Inc.
American Deafness & Rehabilitation Association
American Society for Deaf Children
Convention of American Instructors of the Deaf
Conference of Educational Administrators Serving the Deaf
Deafness Research Foundation
National Association of the Deaf
National Fraternal Society of the Deaf
Self Help for Hard of Hearing People
Telecommunications for the Deaf, Inc.

COR feels the establishment of NIDCD was a great victory for the millions of deaf and hard of hearing people represented and served by our member organizations. But the victory in the battle to establish this new institute has not ended the war to improve the lives of current and future generations of Americans. Much remains to be done.

NIDCD represents a historic opportunity for progress. The Task Force Report indicates a broad range of areas for effective action, and COR fully supports them. However, we want also to ensure and to be assured the NIDCD will have appropriate concern with the behavioral and psychosocial aspects of deafness and hearing loss. This concern was not as obvious in the Task Force Report as some of our member organizations had hoped and expected it to be.

COR's support for the establishment of NIDCD is unqualified. We consider ourselves in common cause and expect to create a partnership with the Institute to address all aspects of deafness and hearing impairment. As a function of this partnership, we ask that every possible step be taken to ensure that a concern for the behavioral and psychosocial aspects of hearing impairment be made an integral part of NIDCD structure and operations.

Our member organizations can individually provide definition as to how this can best be achieved. Clearly, this request is a challenge and a mandate to make NIDCD the best it can be: an institute which comprehensively deals with the understanding, accommodation, treatment and rehabilitation of Americans with hearing and other communication disorders. We trust and hope that this goal is the vision of all who have been associated with NIDCD, and offer the services of COR and our individual member organizations in achieving it.

Sincerely,

Gary W. Olsen
Co-Chair

Donna M. Dickman
Co-Chair

cc: COR Members

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E·A·R
FOUNDATION

April 25, 1989

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National Institutes of Health
Shannon Building, Room 218
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Bethesda, MD 20892

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Dear Mr. Donohue:

Thank you for identifying the Meniere's Network as an organization with an interest in the activities of the new Institute. We appreciate the opportunity to review and comment on "The National Strategic Research Plan" for the National Institute on Deafness and Other Communication Disorders.

The Meniere's Network, a program of The EAR Foundation is a peer support group with patient-organized chapters throughout the United States. The Meniere's Network patients who contact us feel that the opportunity to share experiences and insights with fellow patients is invaluable. The support offered by this Network and its programs is a helpful complement to treatment.

As we talk with these patients we continue to realize the need for a mechanisum such as a national clearinghouse through which these patients can reach the Meniere's Network. We commend your recommendation to establish a planning panel for a clearinghouse (or clearinghouses) at the NIDCD. A representative of the Meniere's Network and/or The EAR Foundation would be pleased to serve on this panel if so invited.

The EAR Foundation was founded in 1971 as a not-for-profit foundation to address the problems of ear related disorders, specifically hearing loss and balance disturbances. From its inception The EAR Foundation has been dedicated to the continuing education of practicing ear specialists, to the development of auditory and vestibular research, and to continued education in these areas, specifically regarding hearing preservation and methods of hearing rehabilitation.

The EAR Foundation offers course programs designed to fulfill the educational needs of ear specialists in practical clinical information and surgical anatomy through observation of televised surgery, temporal bone dissection in the laboratory, and lectures by The Otology Group and leading otologic guest faculty. With this expertise, we would be most happy to assist in any future training program development offered by NIDCD.

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Please feel free to contact me or any members of The EAR Foundation Boards of Directors for additional feedback or assistance.

Sincerely,

Faith Haber Galbraith

Faith Haber Galbraith
Executive Director

GALLAUDET UNIVERSITY

OFFICE OF THE PRESIDENT

(202) 651-5005

KENDALL GREEN
800 FLORIDA AVENUE, N.E.
WASHINGTON, D.C. 20002

April 26, 1989

Dr. Jay Moskowitz, Ph.D.
Acting Director
National Institute on Deafness and
Other Communication Disorders
Shannon Building
National Institutes of Health
Bethesda, MD 20892

Dear Dr. Moskowitz:

I would like to express my enthusiasm for your planning and progress in achieving a National Strategic Research Plan for the National Institute on Deafness and Other Communication Disorders. We at Gallaudet University believe that the creation of the Institute is a milestone that illustrates Congress' desire to expand our knowledge base about deafness and deaf people. Gallaudet pledges to support the NIDCD as it seeks to fulfill this mandate.

I have reviewed the draft of the National Strategic Research Plan Task Force. As you know, members of the Gallaudet research community have been in contact with your Institute with recommendations for clarifying issues of importance to deaf people. In particular, we recommend that the final document cite data on the prevalence of deafness based on currently available government studies. We look forward to the inclusion of this and our other recommendations.

I applaud your efforts to continue to refine your research goals. I also encourage you to interpret your mandate of dealing with health issues in the most comprehensive possible way and to recognize that deafness is a condition that need not be viewed as a communication disorder. Deafness raises many complex communication issues. Some of these require adjustments to normative society, but many others involve recognizing the value of deaf people as people and the importance of natural sign language and the deaf community in their lives. I encourage you strongly to recognize these non-pathological approaches to deafness as you create more specific research agendas based on your recently developed research priorities.

I am proud that three members of the Gallaudet community were included in the Task Force on the National Strategic Research Plan for the NIDCD. We at Gallaudet are particularly pleased that an Advisory Board has been incorporated into the design of the new Institute. It is essential to obtain a variety of perspectives, especially including those of deaf people and professionals whose careers focus on their needs. However, as you know, it has been a matter of some concern that this perspective was underrepresented at the Task Force meetings in January. I am pleased to recognize that since that time you have attempted to obtain the views of a number of organizations and individuals who were not represented in the original Task Force meetings.

It will be our continuing responsibility at Gallaudet to communicate the needs of deaf people to the Institute and to conduct ongoing research relevant to those needs. Gallaudet has been a responsible and significant contributor to research knowledge about deafness and deaf people since 1870, when Edward Allen Fay was doing his work for the American Annals of the Deaf. I have made it a priority for Gallaudet University to continue its preeminence in deafness research.

Once again, I commend you on the progress you and your colleagues have made on creating a national research strategy. Working together, we can guarantee that the new Institute contributes significantly to the betterment of deaf people's lives.

Sincerely,

I. King Jordan, Ph.D.
President

IKJ/cdb

HIA

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April 19, 1989

Mr. Sean Donohue
National Institutes of Health
Shannon Building
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9000 Rockville Pike
Bethesda, MD 20892

Dear Mr. Donohue:

The Hearing Industries Association (HIA), representing the manufacturers and distributors of hearing aids, component parts and related hearing health care products, commends the Task Force for preparing a comprehensive Draft National Strategic Research Plan for the National Institute on Deafness and Other Communication Disorders (NIDCD). The industry is enthusiastic about the purpose and goals of the new NIDCD and wholeheartedly supports a research plan that will benefit not only individuals with hearing and other communication disorders, but also the companies which strive to provide improved technology and assistive instruments to serve those individuals.

Our general comment is that we feel the report strikes a very reasonable balance between short-range and long-range research, and also between the emphasis on prevention an rehabilitation.

Comments on specific proposals are noted below:

1. Reference: Page 1-1, column 2; and Chapter 10, pg. 10-1

Reference in this section is made to the fact that incidence data on communication disorders on a national level does not exist. While the data collection system for information derived from patient population is welcome, we feel the concept of the database should be expanded beyond that of limiting it to incidence data on communication disorders.

At the present time, approximately, 1.4 million hearing aids are sold annually in the United States. Such a large number of fittings could provide the basis for the establishment of a national database which would contain such information on the types of hearing aids sold, the performance properties of the hearing aid, the user's loss characteristics, the typical acoustical environment that the user is exposed to, and some measure of the degree of satisfaction with the prosthesis.

Information of this kind might be obtained with the joint cooperation of the major members of the hearing health care team, i.e., hearing specialists, physicians, audiologists, and manufacturers. Such cooperation might permit the development of mathematical correlations between these factors and user satisfaction, which in turn might provide insight into the efficacy of hearing aids.

2. **Reference: A-15 General Goal, No. 2; and page 2-10 last paragraph**

Report should stress the use of existing research, development and delivery structures to the greatest possible extent. Development of prosthetic devices should be limited to concept testing. Results of such should be publicized and available to all. Institute funds should be exclusively devoted to research rather than commercial development.

3. **Reference: Page 2-6, paragraph 2; and page 2-9, paragraph 9**

Although hearing aids do not restore normal hearing, they are effectively used by millions. Hearing aids undergo continual development and improvement. We feel the report gives inadequate emphasis to the need for research in hearing aids as compared to cochlear and neural implants.

4. **Reference: Page A-17, paragraph 1; and page 8-3, column 2**

Better methods for the evaluation of prosthetic device effectiveness need to be developed. These methods should be based on the recognition of the range of conditions faced in actual use. Much research is needed on stimuli used for testing.

Materials currently used for evaluation include various types of word lists, such as open set, close set, consonant sounds, vowel sounds, sentences, and various background noises. No stimuli standards have been agreed upon. There has been no agreement as to what levels of stimuli should be presented, considering that in the real world, listening occurs in a dynamically changing environment. Additional research in this area is needed.

5. **Reference: A-17, paragraph 2; and page 8-3, column 2**

Research on hearing aid selection procedures needs to involve the consideration of more than theoretical rationalization and computerization of procedures. Research in this area needs to properly validate procedures by evaluating the effectiveness of resulting fittings, and by developing comparisons with fittings resulting from other procedures.

Very substantial, urgent research is needed to further clarify the relationship between hearing loss characteristics and performance characteristics of the hearing prosthesis. A great variety of algorithms have been suggested in the literature for the selection of hearing aids. Some of them are, for example, POGO, NAL, half gain method, Burger (modified half gain) and many others. As a result, hearing aid selection procedures are imprecise, and currently depend much upon user feedback in a process of what is essentially convergent experimentation.

Recent advances in integrated circuit technology give rise to the possibility of incorporating very sophisticated signal processing systems in very small hearing devices. The capability cannot be exploited if research is not substantially intensified relative to the understanding of the relationship between hearing aid performance parameters and hearing loss characteristics.

Additionally, more research is needed on the issue of quantifying efficacy. Two major dimensions describing the efficacy of hearing aids are the perceived sound quality and intelligibility. More research is needed to quantify both of these dimensions, particularly in quantifying perceived quality.

Additional research is needed to quantify the efficacy of a hearing prosthesis in listening situations other than speech, such as for example, music.

HIA strongly believes the Institute would best serve the public by prioritizing hearing research that could be transferred to assistive device production lines. Expanded technology for assistive devices designed for the hearing impaired is critical to the continued development of improved products that benefit more individuals, regardless of their age group, and are acceptable to consumers. Detailed acoustical knowledge and understanding can immediately be implemented to enhance current manufacturing practices, shape response curves, improve hearing testing and hearing aid evaluation, and advance digital technology.

HIA appreciates the opportunity to comment on the NIDCD Draft Research Plan and anticipates working with the Institute on these projects. If we may provide additional information or be of assistance to the Institute in any way, please let us know.

Sincerely,

Carole M. Rogin
Executive Director

T100

NATIONAL HEARING AID SOCIETY

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Rochester, New York
Central East Coast:
ALVIN C. BEISWENGER
Harrisburg, Pennsylvania
Southeast:
HERBERT D. GORLIN
Miami, Florida
Central East:
JON E. DURKIN
Lexington, Kentucky
Southwest:
EDWARD N. RYAN
Baton Rouge, Louisiana
Central:
WILBUR S. MITSDARFFER
LaSalle, Illinois
Northwest:
LARRY B. KENT
Omaha, Nebraska
Pacific:
FLOYD E. LOUPOT
Pasadena, California
Governor-at-Large:
RICHARD L. CLARK
Cleveland Hgts., Ohio

March 28, 1989

Mr. Sean Donohue
National Institutes of Health
Shannon Building, Room 218
9000 Rockville Pike
Bethesda, Maryland 20892

Dear Mr. Donohue:

The National Hearing Aid Society has been contacted by Dr. Jay Moskowitz, Acting Director of the National Institute on Deafness and Other Communication Disorders, and asked to comment upon the draft report entitled "The National Strategic Research Plan for the National Institute on Deafness and Other Communication Disorders".

Initially, we wish to commend the task force of consultants responsible for crafting this draft. Given the diversity of the subject, this was certainly a major undertaking -- yet one which was dispatched in a thorough and timely fashion. While our perspective is admittedly somewhat parochial, the only major comment we will offer is that we would like a greater emphasis placed upon hearing aids as viable rehabilitative treatment.

Again, we appreciate the opportunity to comment and assist in the strategic planning for the National Institute on Deafness and Other Communication Disorders.

Sincerely,

Anthony DiRocco

Anthony DiRocco
Executive Vice-President

AD:rf

SELF HELP FOR HARD OF HEARING PEOPLE, INC.
7800 WISCONSIN AVENUE
BETHESDA, MARYLAND 20814
(301) 657-2248 (V) 2249 (TTY)

April 10, 1989

Mr. Sean Donohue
National Institutes of Health
Shannon Building, Room 218
9000 Rockville Pike
Bethesda, Maryland 20892

Dear Mr. Donohue:

In accordance with a letter from Dr. Moskowitz, comments from SHHH on NIDCD's National Strategic Research Plan are being sent to you.

Our comments are based on the following: 1) a review of the Task Force draft of NIDCD's research plan; 2) a review of the first fifty research projects, by title, listed as now being conducted at NIDCD; and 3) a knowledge of what hearing impaired persons believe is useful to them in the way of research. In preparation for this letter, we have consulted with our members, including Jerome Schein, Ph.D., Laurel Glass, M. D., Ph.D. and Lucille Beck, Ph.D., all of whom are well known to their peers. Dr. Schein is a consultant to SHHH. Dr. Glass and Dr. Beck are members of our Board of Trustees.

We see NIDCD as an institute with an opportunity to break new ground in the area of deafness and hearing impairment. In this context, we were particularly pleased to see recognition, in the Overview on Deafness and Hearing Disorders, that "Even now, there is insufficient public appreciation of the vital importance of good hearing to overall health and employment opportunities" (p. 2-1). In our ten years of work with persons who are hard of hearing we have found the converse to be true as well. Poor hearing can have an adverse impact on wellness and overall health.

The growing interest in the mind/body relationship as it affects the traditional medical model in the United States is pertinent here. If the mind can influence recovery after surgery (we know it can) then perhaps it could also cause the onset of illness (we don't know this with any degree of certainty yet). Depression is one possible source of such linkage. Hearing loss is a fast track to depression. Hard of hearing persons often get stuck in the depression stage of a normal grief pattern after losing a major life sense.

Data suggest that emotional as well as behavioral effects are associated with hearing loss in general and that hearing loss may be especially destructive to the well-being of older persons.

In 1965, 1974, and 1982, approximately 7,000 persons representative of the population of Alameda county, California, were interviewed about their health attitudes and practices. Factor analysis defined five independent assessments in the study: a health attitude index, a health practices index, a social network index, a life satisfaction index, and a depression index (Berkman & Breslow 1983). Recently, the 1965 data were analyzed for the effects of hearing loss on several parameters of well-being. More than twice as many hearing impaired persons were depressed and had low life satisfaction as persons with no difficulty hearing. The proportion of hearing impaired persons who were depressed and had low life satisfaction was markedly higher in persons over 55 years of age.

Such data about the emotional and behavioral accompaniments of age-associated hearing loss are consistent with the writings of professionals working with hearing impaired older adults (Levine, 1960; Luey, 1980; Ramsdell, 1978; Corso, 1977; Oyer et al., 1976), with statements by their family members (Glass, 1986; P. K. Ashley, 1986), with the moving and insightful comments of deafened persons themselves (Elliott, 1978; J. Ashley, 1973; Albert, 1977), and with the limited number of published studies on the psychological and social effects of hearing loss on its sufferer, especially in old age (e.g., Meadow-Orlans, 1986; Kisiel et al., 1984; Myklebust, 1964; Rosen, 1979; Hunter, 1978; Weinstein & Ventry, 1982; Ordy et al., 1979; Orlans & Meadow-Orlans, 1984).

The need for careful research to unravel these biological and psychological complexities and to find ways to alleviate the discomfort the elderly suffer by their hearing loss is apparent.

What this means is that we need more from NIDCD than basic medical research. We must include psychosocial factors. Despite the startling increase in the numbers of hearing impaired persons from middle age to old age, only 12 of 2,445 projects summarized in the Inventory of Federally Supported Research on Aging concerned hearing loss (U. S. DHHS, PHS, NIH, 1982). Of those 12, none dealt with psychosocial implications of hearing loss.

Obviously, the creation of NIDCD should reflect an improvement on that situation. Yet, our examination of the first fifty projects listed on the current projects supported by NIDCD shows nine that may have promise for hard of hearing persons. None include psychosocial implications. Certainly analysis from titles can be misleading and we will examine the abstracts as time permits but clearly NIDCD is being launched without much change in research direction.

NIDCD has the opportunity to bring together, for the first time, a truly comprehensive picture of the medical approach to hearing loss with what complications of hearing loss do to the human being. Remedial directions must be based on the totality of

the two. Hearing loss as an item of medical study is only a small portion of the problem. Today, over 17 million persons suffer from sensorineural hearing loss. It's getting worse. As we search for cell regeneration and other hopeful cures, we must also attempt to alleviate the plight of those suffering with sensorineural hearing loss now. For them, the future is today.

The Surgeon General of the United States has stated that "Mending people, curing them, will no longer be enough. That is only part of the total health care that most people require." Unless this thinking is incorporated into NIDCD's philosophy at an early stage, the Institute's research will have less impact than its potential warrants. In the hearing health field NIDCD could forge the first usable model to provide a basis for the understanding, treatment and rehabilitation of hearing impaired persons, the goal which the American Academy of Otolaryngology-Head and Neck Surgery (AAO) projects as its mandate.

With these background statements, we would like to suggest research which might include:

* The Relationship between Stress and Acquired Hearing Loss in Mature Adults. This research would not only determine the stress correlates of varying types and degrees of hearing impairment, but would also investigate the efficacy of selected stress-reducing tactics and strategies, which might include medicine but would probably emphasize psychological approaches (e.g., joining a self-help group).

* Short- and Long-Term Depressive States in Persons with Acquired Hearing Loss. While similar to the first project, this one addresses another aspect of the hearing loss syndrome. Little empirical research has been done on the course of emotional states and their concomitant (social withdrawal, lability, etc.) following different onsets and degrees of hearing impairment. The research should, like the first, not be limited to descriptive approaches, but should test various means of overcoming depression.

* The Effects of Hearing Impairment on Mortality. To what extent, if any does hearing impairment affect longevity? These epidemiological studies would be directed at uncovering life-threatening consequences of hearing impairment in relation to the age at onset of the disorder, its degree and type, and such other factors as may appear to bear on such a relationship. Depending upon the outcomes of the research, practical steps might be indicated.

Hearing loss is as intrusive as alcoholism. It affects those around the person with the problem. Its generated stress moves both ways--internally and externally to others. Living with

a person with a hearing loss is no easy accomplishment. We need to know:

* The Impact of Hearing Impairment on Unimpaired Family Members. What happens to the families of individuals who suffer hearing impairments? How do spouses and children react when a significant household member loses some degree of hearing ability? The research so far conducted suggests that this is a fruitful area of inquiry, but it has barely scratched the surface.

* Successful and Unsuccessful Coping Strategies of Adults with Acquired Hearing Impairments. What works and what does not work when adults attempt to compensate for loss of hearing? These studies could occupy a fair share of the NIDCD budget...justifiably.

* Employment Prospects Following Acquired Hearing Loss. NIDCD may feel that studies like this belong to the rehabilitation institute, but there are aspects that would warrant joint funding. For example, unilateral impairments are not usually considered for rehabilitation, yet evidence is mounting that losing hearing in one ear has serious consequences that have been overlooked. These effects may significantly impair employment and, therefore, should be given more attention than they have up to now.

In the above, broadly stated areas, both theoretical-scientific and practical approaches are suggested. Our very large constituency of more than 20 million hard of hearing people feels strongly that at least a portion of the research supported by NIDCD have relatively immediate benefits for them. We do not mean to suggest that studies with long-range potential be ignored. Some of the "basic" studies that NIDCD is now supporting may have greater impact on our lives than other more "applied" research. We should not have to choose between them: we should have both and in proportions that provide a large measure of early satisfaction. Not only will that be helpful to hard of hearing people, but also it will be helpful to NIDCD because it will encourage hard of hearing people (some 20 million) to support NIDCD's increasing efforts.

Since we seek research which will enable NIDCD to put together a holistic model of what happens to the human being who loses his or her hearing (in addition to the myriad subjects concerning prevention), here are other areas of interest to hard of hearing people.

Four other directions for intensive research are suggested: (a) biomedical research aimed at the discovery of preventable causes of presbycusis; (b) behavioral science research directed at understanding and ameliorating the handicapping emotional and behavioral responses to hearing loss in middle and later

life; (c) rehabilitation research directed toward development of workable rehabilitation techniques for the communication deficits caused by hearing loss; and (d) technological research to adapt contemporary and future technologies for use by hard of hearing individuals, with emphasis on the needs and limitations of older persons.

In the near future, a small, research-focused, state of the art conference made up of persons active in research and service delivery to adult hearing impaired persons, would be invaluable. Gaps in knowledge and practice about presbycusis could be identified. Feasibilities and priorities for next research emphases could be specified. It might even be possible to set up collaborations so that useless duplication might be avoided but useful replication might be cooperatively defined.

Basic biomedical research might be directed initially toward discovery of the etiologies, results of, and possible treatments for the several origins of presbycusis. Do the repeated episodes of decreased pO2 during the sleep apnea of normal, aging persons adversely affect cochlear hair cells? Which drugs and which drug interactions have permanent effects on hair cell function? Does hair cell degeneration precede loss of auditory nerve cells and nerve fibers in the cochlea, or is the reverse true? Do such degenerations and losses occur independently, or is one causal of the other? Which direction is causality?

Similar kinds of questions are appropriate about Central Nervous System (CNS) components of presbycusis. Where are the CNS lesion(s) association with the slower transmission of auditory stimuli in old persons as contrasted with young? Is such slowing a general characteristic of CNS function in the old or are there components unique to hearing? Is fiber and cell loss in CNS tracts and ganglia greater in the auditory pathways than in other sensory pathways...in sensory pathways than in motor...than in association networks? How does hearing loss affect cognitive function in middle-aged and older persons? Does hearing loss exacerbate the development of confusional states in older adults? What are the effects on cortical function when auditory (and other sensory input) to the arousal centers are decreased or absent?

Behavioral and psychosocial problems, e.g., suspiciousness, depression, low life satisfaction, isolation, stress, confusional states, and so on should be assessed by contemporary research methodologies. Several bodies of longitudinal data include unanalyzed information about hearing loss and its effect on emotions and behavior. Recognizing the constraints introduced by collection methods, by methods of data storage and by the retrospective nature of the analysis, it seems important to retrieve the extensive information buried in these longitudinal data, accessible at relatively low costs. Their systematic analysis can give significant information about correlations

between hearing loss and occupation, geographic region, diet, racial and ethnic group, and so on. Insight from such studies can be used to direct the design of prospective research focused on specific aspects of affective and behavioral effects of hearing impairment in older persons. In particular, new instruments to assess psychosocial function in hearing impaired persons should be developed and validated on adult populations. Note that instruments designed for and tested with college students are not appropriate for uncritical use with middle-aged and old persons.

Ototoxic drugs is another area for continued research. The effects of the aminoglycosides are known to most physicians who therefore are commendably cautious in their use. However, too little is known about the effects of other drugs that frequently are prescribed for older persons. Furosemide (brand name Lasix), for example, is the diuretic prescribed most often for persons 65 and older and the third most frequently prescribed diuretic for persons between 45 and 64 years of age (Cypress, 1983). What most physicians, even geriatricians, don't know is that furosemide may cause hearing problems that are irreversible when the drug is administered chronically and at high dose (Rybak, 1982). More research is needed to clarify the range of doses and chronicity at which furosemide is ototoxic and to identify which patients are most susceptible to furosemide ototoxicity. Nor is it known whether furosemide interacts with other drugs used frequently with older patients to increase damage to the auditory system.

As little as is known about furosemide, even less is known about direct and/or interactive effects on hearing (or cognition or affect or the feeling of well-being, etc.) of the mutiple drugs many old people are told to use daily. These are issues of major importance, not only for persons already old but also for the middle-aged who are moving daily closer to the health problems often associated with old age.

Rehabilitative methodologies are difficult to study, primarily because personal style--both of the rehabilitation professional and of the hearing impaired individual--are so intimately involved in outcome. Nonetheless, efforts to supplement classical rehabilitation techniques with supplemental skills, e.g., simplified manual languages such as a simplified Signed English or a simplified cued speech, might add significantly to rehabilitation success. Research into the reasons why so many older persons refuse to use hearing aids is imperative.

Perhaps a major problem that contributes to hearing loss being low on the concern scale of even those who have it, is the lack of understanding of its full complications by professionals who are often the origin of attitudes held by lay people in matters of health. Studies in pharmacology have already indicated that the attitude of the prescribing caregiver may have even

more impact than the prescription itself. Hearing Industries Association (HIA) contracted research touches on this subject in regard to hearing health. We need research on attitudes and referral practices of hearing health care providers towards persons of varying ages, with comparable losses of hearing and variations in recommentations for follow-up care, rehabilitation and utilization of assistive devices, including hearing aids. For example, a 30-year old person with a mild to moderate hearing loss will normally be referred for care/rehabilitation while a 70-year old person in the same circumstances may not be referred. Does the attitude that "you hear pretty well for your age" which establishes a reduced level of expectation (unnecessary) start with the care giver or the hearing impaired person?

Nguyen (1984) studied denial in older persons who were found to be hearing-impaired by a screening program. She found, unexpectedly, that their attitude toward the hearing loss had no significant correlation with whether or not they followed through on referral to a physician. Equally unexpected was the finding that the audiologist's failure to refer was of primary significance in determining whether or not the individual received further workup. Rehabilitation Services Administration data also suggests that these problems deserve further examination.

Perhaps because of the association with getting old and with the negative image old age has had in our society, perhaps because of the threat of increasing isolation implicit in hearing loss, and perhaps because of the common assumption that hearing loss and low function go together, old persons as well as others often deny that they have a hearing problem. The reasons for denial are complex and not well defined by research. Whatever its cause, denial makes diagnosis and treatment of the hearing impairment more difficult.

In SHHH, we have seen the consequences of this to be profound. Such attitudes restrict progress in all areas of hearing health and will reduce the impact of NIDCD's research unless understood and corrected.
Technological research should continue to be directed toward making hearing aids more manageable and tolerable particularly by older persons who are the most frequent users. In particular, an inexpensive hearing aid is needed which can be operated simply and repaired easily. Procedures for the inexpensive provisions of custom-made ear molds for hearing aids are also needed. Continued efforts to develop other assistive devices and alternative listening equipment are needed, with emphasis on reasonable cost as well as on electronic state of the art. Creative thinking on the use of computers and new types of telephone transmission as well as the involvement of other senses in the communications process are needed. Hearing is an important but only one aspect of communication.

A word about the clearinghouse. Hearing loss is not benign.

Though of high prevalence, hearing loss is of low concern among most health care professionals, and among our society in general. Ignorance and the attitudes it breeds are major forces inhibiting progress in the field of hearing loss. And yet, as hearing impairment strikes at the very essence of being human, it impacts in a myriad of ways. It restricts the ability to be productive--frequently manifested in lower career expectations, lost jobs or early forced retirement; it limits social intercourse; it has a major impact on relationships with spouses and family members; it reduces constructive use of leisure time; it affects physical and mental health; it often leads to poor self-image, to isolation, to despair and ultimately, it can affect the will to live. NIDCD's National Information Clearinghouse could help change the perceptions and attitude of both health professionals and the general public by dissemination of accurate information about all facets of deafness and hearing impairment.

Less frequent causes of decreased hearing in the old may be the late-acting results of viral diseases in infancy or childhood, or even of late-appearing developmental dysfunctions. Since avoidance of preventable hearing loss in later years is related primarily to protective measures taken in earlier ones, it is clear that individuals of all ages need education about how the auditory system works. Only with such understanding can they realize the things they must do if they are to avoid damaged hearing later in life.

Existing networks and organizations have much to offer, but the challenge to accurate information begins with the different terminology used by different groups when discussing the same thing. Review of existing literature for use in a more standardized way will be necessary. The idea of a panel to help define the scope of activity and develop guidelines for the clearinghouse is excellent--but <u>all</u> subpopulations of hearing impaired persons must be represented.

Finally, while we understand NIH's emphasis on training the physician to be an investigator, NIDCD has the opportunity to lead the way, and in the process to attract more personnel, by training people with research, rehabilitative and technological emphasis. Biomedical engineering personnel comes to mind.

Mr. Donohue, we know NIDCD's limitations and constraints. Our comments are offered in the context of awareness of consumer interest, potential for NIDCD, and an acute need to redress the balance of research to include some of the subjects discussed in this letter. Judicious use of NIDCD's funds based on a philosophy

that the whole person must be considered in hearing health, could achieve what most of us who are taxpayers, as well as hearing impaired persons, would like to see from the NIDCD.

Thank you for this opportunity to share our thoughts with you. I would like to acknowledge again the significant input to this letter from Dr. Schein, Dr. Glass and Dr. Beck.

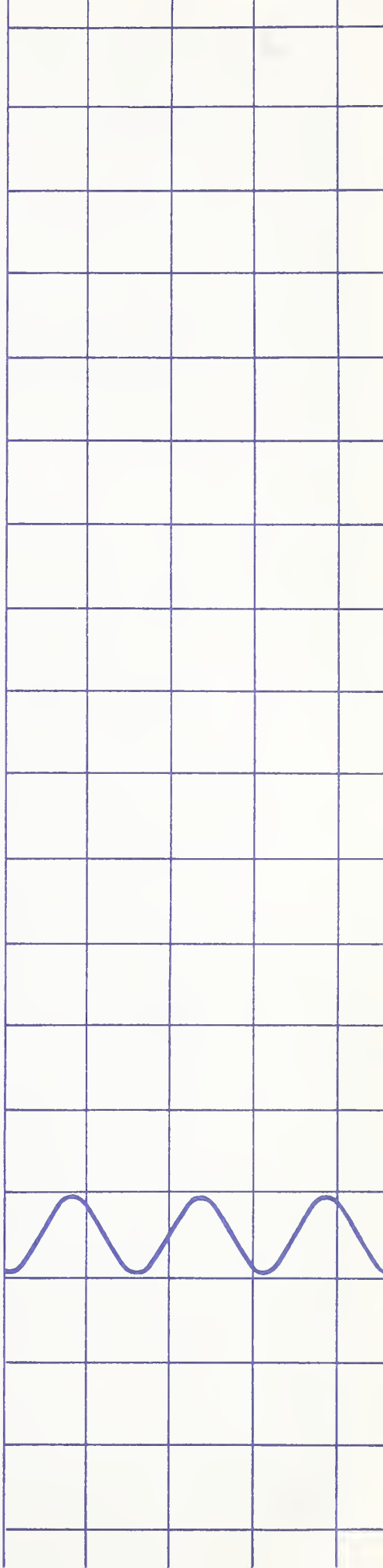
Sincerely,

Howard E. Stone

Howard E. Stone, Sr.
Executive Director
mhg

Appendix E.

A Comprehensive Program of Research on Deafness and Hearing Disorders



A COMPREHENSIVE PROGRAM OF RESEARCH ON DEAFNESS AND HEARING DISORDERS

INTRAMURAL PROGRAM

The existing intramural program of the NIDCD consists of the Laboratory of Molecular Otology, a speech and voice unit, an audiology service, and an otolaryngology-head and neck surgery service.

The Laboratory of Molecular Otology was established in 1974 as the Laboratory of Neuro-Otolaryngology in the NINCDS intramural program. Its mission is to perform multidisciplinary research on the basic mechanisms of normal hearing. The present laboratory consists of three senior scientists with expertise in the area of biochemistry, cell biology, and immunobiology. With a supporting staff of 11-14, the laboratory provides leadership in introducing many new innovative techniques to the field of auditory research, including the use of immunohistochemistry to characterize the neural organization of the cochlea and the auditory nuclei in the brain stem. The laboratory is a leader in the purification and characterization of a neurotransmitter receptor that may be critical for the function of the auditory sensory cells and the auditory nerve. The laboratory has successfully produced monoclonal antibodies against cochlear cells. It is also a leader in the area of advanced morphology combining light microscopic, immunohistochemical, electron microscopic, and cryobiological techniques to characterize the molecular organization of the cochlea.

The speech and voice unit was established in 1984 in the NINCDS intramural program. Its mission is to conduct research on normal laryngeal and vocal tract structure and function and to improve understanding of speech and voice disorders, which will lead to improved prevention, diagnosis, and treatment. The present staff comprises one senior scientist and four additional scientists with expertise in speech and voice pathology, otolaryngology, and neurophysiology. The program provides national and international leadership in the pathophysiology and treatment of spasmodic dysphonia, dysarthria, and stuttering.

The audiology unit has been in existence since 1979 and is the primary source of clinical service and clinical research in audiology at the NIH. It collaborates with a broad range of programs and institutes, including particularly close liaison with the interinstitute genetics program. Research performed by the audiology unit is leading the discovery and analysis of human hearing dysfunction in diverse genetic, neurodegenerative, and metabolic disease processes. Recent collaborative efforts in applying principles of Artificial Intelligence to audiology have resulted in the development of a PC-based expert system for use in ABR evaluations. Vestibular study has recently been added to the intramural audiology operation.

The unit is currently staffed by its founder (and chief) and three other professionals. It also serves as a training site for graduate students in audiology and otology.

The otolaryngology-head and neck surgery service provides services to the Clinical Center and plans to become involved in clinical research related to reconstruction of the vocal tract and nasal physiology.

The present intramural resources in terms of program diversity, personnel resources, and laboratory space are inadequate for meeting the mandate of P.L. 100-553. These resources must be expanded. While it is recognized that the intramural program cannot encompass the entire scope of communication disorders, there is a pressing need to introduce new programs with great potential for yielding novel and critical information. Such new programs must include molecular biology, molecular genetics, advanced cell physiology, and electrophysiology applied to the auditory system in animal models and in humans. An expanded intramural program could serve as a central gene bank for the emerging research on the genetics of deafness.

There is a very pressing need to expand intramural clinical research, utilizing the existing audiology and speech units and the otolaryngology-head and neck service as a base. New programs must be added to address more types of hearing disorders, to coordinate multicenter clinical trials, to perform research on human hereditary deafness, and perform studies of the molecular mechanisms of hearing and deafness.

EXTRAMURAL PROGRAM

Normal Hearing

A. ***Determine how the ear processes and encodes sound***

The ear can be divided into three main sections:

- The external ear, which includes the part of the ear visible to others and extends to the ear drum;
- The middle ear, which is the part between the ear drum and the inner ear; and
- The inner ear, where the actual transduction of sound into nerve impulses occurs.

The outer, middle, and inner ear work together to transduce acoustic information into patterns of nerve impulses. Research on transduction has brought investigators to the verge of a complete description of the physiological and molecular mechanisms of the transduction process. The following outline gives details of experimental approaches that will yield additional essential information in the near future.

1. *Outer and middle ear*

   The function of the external ear in affecting an animal's or person's auditory perception of space is still not completely understood and needs to be further investigated, particularly in regard to how movements of the external ears of animals are integrated into the process.

   Sound conduction within the middle ear is relatively well understood, but more information is needed on the role of the middle ear muscles and their control by the brain. In addition, the mechanisms by which the cells of the middle ear maintain their function in the face of various noxious agents such as bacteria need further elucidation.

2. *Inner Ear*

   The inner ear is the site where mechanical stimuli are transduced into nerve impulses. While this process is complex, three well-defined areas of concern can be identified:

   a. *The control of mechanical vibrations in the cochlea*

      - Establish the biophysics of the active processes in the inner ear and the physiological control of the mechanisms underlying the motility of hair cells.
      - Define the origins and the underlying mechanisms of otacoustic emissions; and investigate how these emissions relate to the normal and abnormal functioning of the inner ear.
      - Incorporate recent advances in knowledge into models and hypotheses of cochlear function that will guide in developing experimental design.

   b. *The mechanisms of the transduction of mechanical vibrations into nerve impulses*

      - Investigate the molecular basis of the mechano-electrical coupling in the sensory cells.
      - Establish the molecular mechanisms of the synaptic processes that are triggered by the acoustic stimulation and that communicate the information to the auditory nerve.
      - Elucidate the way in which the complex information in auditory signals (intensity, frequency, and temporal characteristics) are encoded into information carried by the auditory nerve.

- Elucidate the mechanisms by which the central nervous system influences processing in the cochlea.
- Clarify the relationship between the neural codes of intensity, frequency, and temporal characteristics and the perception of these variables.

c. ***The mechanisms that maintain homeostasis in the inner ear***

All processing of acoustic information requires a strictly maintained environment in the inner ear. Dysfunction of its control mechanisms leads to hearing dysfunction.

- Investigate the mechanisms by which inner ear fluids are generated and maintained.
- Elucidate the mechanisms that regulate blood flow, oxygen, and nutrient supply in the inner ear.
- Investigate the immunological mechanisms that may be operating within cochlear structures.

B. ***Determine how the central nervous system processes the output from the ear***

A primary goal of research on the inner ear is to achieve a detailed understanding of the nerve-impulse code that is sent to the brain after any sound, particularly speech, is transduced. The organization and function of the central connections relative to hearing function, feedback system, and molecular biology of the central nervous system must be studied. This information, which has only partially been determined, is essential to a number of areas of both basic and clinical investigation. First, scientists studying auditory pathways in the brain need to know the nerve-impulse code. Second, language scientists need to know the code, because they derive from it those aspects of the information contained in sound waves that are actually transmitted and thus available to the brain. Third, designers of hearing aids and cochlear implants need to know the code, because, with most of these devices, that is the exact information they are trying to replace in the auditory nerve of an individual with impaired transduction.

1. ***Study at the molecular, single cell, and neural network levels of organization the ways in which auditory nerve-impulse signals are processed in the auditory system of the brain***

Particular attention should be given to neural representation and processing of frequency, intensity time, location of a sound source in space, and complex sounds including speech and other biologically relevant stimuli.

2. *Establish the role of efferent nerve fibers innervating the cochlea*

   Neural signals travel not only from the ear to the brain and from lower to higher brain centers, but in the reverse direction. The understanding of such "descending" or "efferent" inputs to the cochlea is advancing rapidly and may hold a key to understanding the effects of noise on hearing.

3. *Determine the nature of synaptic inputs along the auditory pathway*

   Signals are transmitted from one neuron to another via connections called synapses. The signal processing properties of single neurons are in part determined by the spatial pattern of excitatory and inhibitory synaptic inputs to the neuron. Understanding these patterns of inputs and synaptic mechanisms is crucial to an understanding of auditory signal processing in the brain.

4. *Investigate the mechanisms of plasticity in the auditory system*

   Synaptic connections and, therefore, sound representations in the auditory system can be influenced by learning, experience, and injury. Study of such plasticity leads to insights into the neural mechanisms of auditory signal processing and is directly relevant to a clinical understanding of the effects of lesions in the auditory system.

5. *Establish the cellular organization of the auditory system in the brain*

   The gross structures of the auditory system in the brain and their interconnections have been known for some time. New techniques, however, enable discovery of the connections between individual neurons in one brain center with individual neurons in other structures. Tracing of connections with such precision is allowing rapid progress in understanding the organization of auditory systems. Coupled with physiological recordings from the same cells, such neuroanatomical studies are allowing the generation and testing of hypotheses regarding the organization of neural systems for auditory signal processing.

6. *Extend knowledge of auditory space perception in man*

   Sighting of a sound source in space involves the coordination of visual and auditory senses and motor control. Studies of the interaction of visual and auditory systems in the barn owl to produce a map of sound source location is one of the most exciting developments in hearing science in the past decade. Similar studies are needed for humans.

7. *Establish functions of parallel signal pathways from the cochlea to the cerebral cortex*

   There are a number of signal pathways from the cochlea to the cerebral cortex. Their functions are of primary importance in hearing science. For example, in the barn owl, there is new evidence that time and intensity aspects of a sound may be processed by different brain stem systems and combined to produce a space map at higher levels in the brain.

C. *Determine how central nervous system processing leads to perception and behavior*

Sounds are transduced into a nerve-impulse code in the cochlea. Complex sounds consist of spectral and temporal components, and the code must include some or all of this spectral and temporal information. Perception of the meaningful sounds of everyday life is mediated by the auditory system in the brain. The brain must process this complex neural code in order for auditory perception to occur.

1. *Research questions*

   a. *Perception of acoustic space.* In humans, the ability to localize a sound's source is based largely on sound inputs to both ears. The neural fibers from each ear converge within auditory centers in the brain. How the brain processes this binaural information to account for the perception of auditory space is a topic of very active investigation. Recently developed methods for presenting sounds to subjects over headphones in a way that they are perceived as if they had occurred in auditory space provide a valuable tool for studying localization. This signal processing technique may also be useful for hearing aids.

   b. *Perception of complex patterns.* Because complex sounds consist of many spectral and temporal components, understanding how animals and humans process and perceive complex patterns of these components have revealed important insights concerning auditory perception. Of special importance are the mechanisms by which neural codes for the spectral and temporal components interact when a complex sound is presented.

   c. *Specialized animal systems.* Sound localization by owls, echolocation by bats, song recognition by certain birds, sensory reception by fish lateral line organs, and communication by weakly electric fish are some of the specialized sensory processes that, through research, will continue to provide valuable and exciting insights about auditory processing of complex sounds.

d. ***Acoustic basis of speech perception.*** Speech perception depends on a number of processes including hearing, learning, and language. Understanding how the auditory system processes the neural codes associated with the components of speech will provide a crucial contribution to a full understanding of speech perception.

e. ***Selective auditory attention.*** Most everyday listening situations involve a number of sound sources, many of which are attended to at the same time. One is able to attend to these multiple sources despite the fact that the auditory system receives only one complex sound from each ear. Understanding how the auditory system sorts a complex sound field into its constituent sources is important for understanding normal auditory perception and, possibly, for helping the hearing impaired person process sounds in complex multisource environments.

f. ***The relationship between the cochlear code for sound and central nervous system factors.*** Important new contributions will be made by relating the code for intensity, frequency, and time to the perceptions of pitch, loudness, timbre, and dimensions of complex sound.

2. *Technique*

a. Additional development of noninvasive physiological techniques such as positron emission tomography (PET) and continued use of new analytic and computer models will help to tie knowledge of neural function to that of perception.

b. By comparing the auditory capabilities of animals across the animal kingdom, much has been learned about auditory processing. This comparative approach offers additional promise in the future and remains a valuable technique for studying auditory processing.

D. ***Characterize the life cycle of the normal auditory system (development, maturation, and aging)***

The development and maturation of normal hearing, the auditory problems of children, and changes in hearing that often accompany aging are special phenomena pertinent to the life cycle of the normal auditory system. To date, there have been isolated studies of particular developmental and aging phenomena. The results have shown that there is great interdependence in the development and maintenance of normal auditory system function, so a more broadly directed approach is required to

gain the basic understanding necessary to prevent, diagnose, and treat disorders associated with development and aging.

1. *Determine the molecular genetic basis of normal and abnormal auditory structure and function, particularly as it pertains to developing and aging systems*

   These efforts should include the development of gene expression (cDNA) libraries for the major cell types in the cochlea in mice and humans.

2. *Characterize the normal developmental mechanisms that lead to the formation of the ear and the auditory pathways in the brain*

   This information is essential for understanding abnormal mechanisms that produce congenital defects in children and for efforts to correct them.

3. *Investigate changes in plasticity and adaptability in both the ear and the brain*

   a. Evaluate changes in the strength of interactions and interdependencies in the developing auditory system. The nature and timing of such interactions are likely to govern when the individual is susceptible to a compromise of hearing function.
   b. Evaluate influences of environmental factors through several stages of auditory development, maturation, and aging.
   c. Investigate metabolic, biochemical, and immunological changes that occur during the life cycle of the normal auditory system in the middle ear, in the cochlea, and in the brain.

4. *Determine what mechanisms underlie sensory cell regeneration and recovery of hearing in damaged auditory systems in animals that readily recover from "nerve" deafness*

   Success in this research has the potential to lead to self-repair mechanism for millions of children and aged individuals who are affected by currently irreversible "nerve" deafness. Recent discoveries in animals have demonstrated the existence of an auditory repair process analogous to the familiar healing of wounds in skin. The challenge is to transfer phenomena identified in animals into the ears of humans. Investigations should combine utilization of tissues and cells maintained in culture with studies in animals, where regeneration of sensory cells has been demonstrated or is suspected.

5. *Identify and characterize normal aging processes as they affect the auditory system to better understand abnormal effects in aging*

   Psychoacoustic and speech perception population studies and studies of aging anatomy and aging physiology in animal models and in humans are essential to answer the question: What is normal aging in the auditory system? Any study of aging in human populations must recognize that "normal aging" is only a theoretical construct. In the real world, environmental factors in particular modify the aging process in ways which vary across societies and over time. For example, if smoking and noise exposure decline, the amount of hearing loss seen in "normal" aging populations may also decline.

6. *Accelerate progress toward an understanding of developmental mechanisms and changes in the aging auditory system by capitalizing on advances in biotechnology*

   This effort will entail devising more and better in vitro, molecular, immunological, and optical techniques.

E. *Acquire information and techniques from other disciplines*

The interdisciplinary nature of research on deafness and other communication disorders is both a major strength and a challenge. The strength lies in the accumulation of information on a single sensory system from the conceptual and experimental strategies and technological advances of many different basic sciences. The challenge lies in integrating the information, coordinating experimental approaches, and maintaining open communication between the various disciplines. To maintain and foster the rapid rate of progress in auditory research, a better integration of existing research areas is needed, along with the use of new technologies developed in other disciplines. The following should be incorporated into experimental designs:

1. Recent advances in computer and cognitive sciences related to digital signal processing, parallel processing, artificial intelligence, and neural networks.
2. Technological breakthroughs in laser, optical, and imaging technology, such as confocal microscopy/laser fluorometry for optical analyses of tissues; and magnetic resonance imaging (MRI) and positron emission tomography (PET) for noninvasive studies of the brain.
3. The emerging tools of molecular genetics, molecular and cellular biology, and immunology, including techniques of gene mapping, application of molecular probes, specific immunocytochemistry, and development of targeted monoclonal antibodies.

4. Newly developed multichannel electrodes for simultaneous recordings from multiple neurons.
5. In vitro model systems for study of specific mechanisms of inner ear physiology and function in a controlled environment. Such models might include slice preparations from the central nervous system, isolated cochlear tissues such as the organ of Corti or stria vascularis, as well as primary cultures of neurons and hair cells.

Hearing Disorders

Multidisciplinary studies focused on a number of specific hearing disorders about which little is known are needed. Specific disorders are discussed below in terms of what is known about their prevalence, etiology, pathogenesis, diagnosis, treatment, and prevention.

Acoustic Tumors

Epidemiological studies need to be instituted to define the prevalence of acoustic tumors, of which there are several types, including the bilateral form (neurofibromatosis, type 2) as well as the classical unilateral acoustic neuroma. Genetic studies, including identification of markers and gene mapping, should be undertaken. Methods for early detection need to be refined, including use of ophthalmologic correlates, radiographic imaging, and auditory electrophysiologic techniques.

A comparison of surgical and medical management strategies should be investigated for each type of acoustic tumor. Development and use of neural prostheses in these populations of adventitiously profoundly deafened people should be investigated. Methods to identify and educate at-risk populations need to be developed for counselling high-risk patients.

"Autoimmune" Ear Disease

A group of inner ear disorders is characterized by rapidly progressing sensorineural hearing loss. Recovery of hearing often occurs, when patients are given steroids and/or cytotoxic drugs. The etiology and pathogenesis of the disorder(s) are not well understood; an autoimmune mechanism has been postulated.

Before epidemiological and natural history data can be collected, there is an urgent need to establish a working definition of the disorder(s) that incorporates consistent clinical and immunological profiles. The disorder(s) can occur independently or in association with known systemic autoimmune disorders such as Cogan's syndrome, Crohn's disease, and Wegener's granulomatosis. There is a need to characterize the immune mechanisms affecting these patients. Appropriate animal models should be developed to establish the pathogenetic mechanisms.

An effort should be made to establish acceptable diagnostic criteria and to develop more specific tools to improve the accuracy of the diagnosis.

Improved methods of managing patients with these disorders are needed.

Genetic Hearing Loss

A genetic cause or defect is suspected to account for approximately 50 percent of both congenital and late-onset hearing loss. To date, very little research has been focused on genetic hearing loss. There is an immediate need to locate and characterize the gene or genes responsible for deafness, in both humans and known animal models of genetic deafness. Results may lead to a possibility for the prevention and treatment of the disease by biochemical means.

There is also a need for population studies to identify the epidemiology of genetic deafness.

The prevention of genetic hearing loss will become possible through genetic counseling.

Infectious Diseases

There are many infectious disorders that can cause hearing loss. The leading cause of acquired sensorineural hearing loss in children is bacterial meningitis. Otitis media causes widespread, usually temporary, conductive hearing loss in children. Other infectious disorders known to cause hearing loss include viral disease, such as cytomegalovirus infection, rubella, and mumps. Syphilis also can cause hearing loss.

There is a need to better understand how infectious agents actually cause hearing loss by finding the microscopic and ultrastructure locus of damage and by determining how infectious agents adversely affect embryogenesis when a prenatal infection has occurred. In addition, further research is needed to:

- Determine the incidence of severe hearing loss resulting from bacterial meningitis, and document the natural history of the hearing loss once it has developed. How soon after meningitis does the hearing loss become manifest? Does the hearing loss remain stable, improve, or worsen?
- Develop more rapid and more specific diagnostic tests than are presently available to determine the degree to which inner ear structures are affected by infectious agents. Improved serologic, immunoabsorbent, and audiologic tests are needed.
- Develop therapeutic agents effective in "protecting" the integrity of the inner ear in patients who have infectious disorders known to be highly correlated with deafness. Possibly, antibodies can be developed that will bind to inner ear structures and prevent the deleterious effects of infection.
- Test the efficacy of vaccines in preventing otitis media.

Meniere's Syndrome

Meniere's syndrome is a common cause of unilateral fluctuating sensorineural hearing loss. From data derived from European population studies, the incidence of bilateral hearing loss has been estimated as occurring in 5 percent to 20 percent of the cases. Similar epidemiologic data are not available for the heterogeneous population of the United States. These studies should be initiated.

The cause of the hearing loss in Meniere's syndrome is not known. There is a need to determine whether immune pathologic processes, infectious agents, or environmental agents play a role in the pathogenesis of this disorder.

The pathophysiologic processes responsible for inducing the fluctuating hearing loss are unknown. Research in the area of fluid dynamics, ion transport, and cochlear hair cell mechanics will contribute to the understanding of the mechanisms underlying the resulting hearing loss.

The diagnosis of hearing loss in Meniere's syndrome has relied on traditional audiologic measures. These methods have not been sensitive enough to predict the efficacy of treatment. Newer diagnostic tools need to be explored, based on electrocochleography in otacoustic emissions and psychoacoustic tuning curves.

The treatment of Meniere's syndrome has been both medical and surgical. The emphasis of treatment has been on control of the vestibular symptoms of the disorder. There have been sporadic reports that the hearing loss can also be controlled. Prospective clinical trials to establish the efficacy of these treatment modalities should be initiated.

Tinnitus

The perception of a sound (ringing, buzzing, or roaring) when no external acoustic sound is present is called tinnitus. Millions of people suffer from tinnitus and some have their lives completely disrupted by their severe tinnitus. Essentially nothing is known about the prevalence or cause(s) of tinnitus. Research is needed to:

- Document the prevalence of tinnitus and its relation to other forms of hearing disorders.
- Develop an animal model for tinnitus.
- Better understand the relationship between aspirin toxicity and tinnitus.
- Develop effective clinical measures of tinnitus sensation and disability.
- Continue to understand the psychophysical consequences of tinnitus.

Otosclerosis

Otosclerosis is a chronic disorder of the bony labyrinth and stapes, causing progressive hearing loss. There is a known genetic predisposition for the disease.

Although the pathology of otosclerosis is well described, its pathogenetic mechanisms need to be elucidated. Among the suspected causes are virus, immunologic mechanism, and inborn errors of metabolism affecting bone or connective tissue cells.

- Recent advance in the areas of gene mapping, HLA typing, cellular and molecular biology, cell culture techniques, and immunobiology provide tools to address these questions on a new level.
- The epidemiology of cochlear otosclerosis should be determined. The pathological correlates of cochlear otosclerosis should also be documented.
- There exists an opportunity to improve methods for early and accurate diagnosis of otosclerosis, particularly cochlear otosclerosis. Promising methods include improved imaging techniques, genetic markers, and immunological techniques.
- There is a need to evaluate the efficacy and cost effectiveness of various treatment modalities. New innovative pharmacologically based methods of treatment are needed particularly for cochlear otosclerosis.
- An effort should be made to identify high-risk individuals and patients with a confirmed pathological diagnosis after middle ear surgery, who may benefit from prophylactic therapy for cochlear otosclerosis.
- The ultimate goal should be prevention of all forms of otosclerosis by early detection and preventive measures.

Perilymph Fistula

Leaks of perilymph from the oval and round window areas may occur after barotrauma, after head injury, or spontaneously, and may cause both hearing loss and balance problems. Perilymph fistulas (PLFs) are often difficult to diagnose; even surgical exploration of the middle ear may be inconclusive. In the absence of a valid standard for diagnosis, it is difficult to study the prevalence of this disorder or to assess the accuracy of noninvasive tests for PLF. An important immediate need is for studies of intraoperative diagnosis of PLF. Do otologists agree on intra-operative observations? If not, can a test be devised to document the presence of perilymph in the middle ear?

The latter question—and several others—can best be pursued initially through animal models of PLF. For example, how do PLFs cause hearing loss and dizziness? Does morbidity from PLF depend on interactions with other factors such as pressure fluctuations, noise, or patency of cerebral spinal fluid-perilymph connections? Does intervention

(restriction of activity or surgical closure) at different stages of disease reverse or stabilize morbidity? Are healed PLFs more susceptible to subsequent barotrauma? Can noninvasive tests (for example, involving ear canal pressure changes) detect open PLFs? Can these tests distinguish open PLFs from closed PLFs, hydrops, or vestibulofibrosis?

If clinical diagnosis can be improved, studies of prevalence, noninvasive tests, and therapy should have high priority.

Sensorineural Hearing Loss

Sensorineural hearing loss or so-called "nerve" deafness is the most common form of hearing loss, affecting more than 17 million Americans, 80 percent of whom have significant hearing loss. "Nerve" deafness has been considered a permanent condition. Because the production of sensory cells in the ears of mammals ceases before birth, damage later in life was thought to be irreparable. The recent reported potential for sensory cell regeneration is exciting and must be thoroughly investigated.

The potential development of improved hearing aids that can permit high gain amplification needed to mitigate against the decreased sensitivity of the "nerve" deaf ear is being pursued in several laboratories and should continue.

In addition, it is important to investigate the etiology of sensorineural deafness in order to determine the nature and origins of the classes of disorders that are grouped in this broad category. New directions for therapy should be sought based on the location of the lesion, its extent, and its origin. The potential benefit is unusually great.

Sudden Deafness

Sudden hearing loss is a devastating disorder. Little is known about the etiology, incidence, or pathophysiology of sudden deafness. Even the definition of "sudden deafness" is not generally agreed upon. There is a need to clearly define the disorder and to convene a task force of acknowledged experts who can agree on terminology and criteria to be used for diagnosis. At present, numerous medical and surgical therapies are being used to treat sudden hearing loss or deafness, and no single therapy has been proven to be effective.

Research is needed to:

- Determine the lifestyle risk factors and congenital anomalies that may predispose patients to develop sudden deafness.
- Determine the statistical probability for spontaneous recovery versus the predictability of worsening hearing without medical or surgical intervention.
- Collect and analyze temporal bones from patients who had suffered from sudden deafness.

- Determine the cost:benefit of radiographic, serologic, endocrine, and metabolic tests often requested for diagnosis of the etiology of sudden deafness.
- Develop clinical trials comparing efficacy of such therapeutic methods as: bedrest, carbogen, hemodilution, steroids, volume expanders, anticoagulants, viscosity altering agents, surgical repair of perilymph fistula, and endolymphatic sac surgery. To accomplish meaningful studies, large numbers of patients will be needed. Such efforts should be possible through multicenter controlled clinical trials.

Rehabilitation of Hearing Disorders

In spite of recent and anticipated progress, the ability to prevent or cure hearing disorders will remain less than perfect for many years to come. A major thrust of research will, therefore, be on habilitative and rehabilitative management. This work has three principal goals. More detailed information on these three topics follows.

General Program Goals

- Develop a comprehensive description of the characteristics and consequences of hearing disorders.
- Develop a range of prosthetic devices (hearing aids, cochlear implants, and tactile aids) that will restore adequate sound and speech perception to all hearing impaired individuals.
- Develop behavioral approaches to educational and communicative intervention that will appropriately minimize the consequences of impaired reception.

A. *Consequences of Hearing Disorders*

An important goal is to delineate the consequences of hearing disorders that remain after the possibilities for curative treatment have been exhausted. Work in this area could include:

1. ***Psychophysical studies of the disordered ear, of the aging ear, and of the electrically stimulated ear***

   Studies of tactile and visual psychophysics might also be included to the extent that this impinges on the development of sensory aids, speech training aids, prosthetic devices, and rehabilitative management.

2. *Speech perception studies of the disordered ear*

   As with psychophysical studies, this work could include electrical, tactile, and visual speech perception to the extent that they impinge on prosthetic and rehabilitative management.

3. ***Studies of the linguistic, educational, social, emotional, and vocational consequences of hearing loss, of the mechanisms by which those consequences occur, and of the factors that interact with hearing loss in determining their magnitude***

4. ***Studies of auditory processing disorders that do not necessarily cause changes in auditory threshold (auditory agnosia/dysgeusia)***

   Such difficulties often go unrecognized by professionals concerned with hearing disorders and yet may be a major contributor to learning difficulties in children and to comprehension difficulties in neurologically compromised adults and the elderly.

5. ***The development of assessment methods to permit accurate description of persons affected by hearing disorders, to contribute to determination of cause, to contribute to the selection of prosthetic and rehabilitative management options, and to aid in the comparative evaluation of various interventions***

Identified priorities in this area are:

- Delineation of the psychoacoustics of sensorineural hearing loss, the dependence of psychoacoustics on causation and pathology, and its influence on auditory speech perception.
- An understanding of the nature and mechanisms by which recurrent otitis media in children leads to long-lasting difficulties of perceptual function, language function, and/or learning style.
- Delineation of the special problems encountered by hard-of-hearing children, including their perceptual characteristics, their ability to deal with noisy and reverberant surroundings, the difficulties encountered in speech and language acquisition and educational attainment, and the socioemotional consequences of their hearing difficulties.

B. *Hearing Aids and Other Prosthetic Devices*

Many diseases of the auditory system result in disorders of communication as a result of reduced sensitivity and resolution for sound. Presently available prosthetic devices for such impairments include hearing aids, cochlear implants, and tactile aids.

Significant progress has been made in the last decade in improving and evaluating the effectiveness of these devices, and there is substantial promise that these advances will continue.

Conventional hearing aids attempt to overcome, through sound amplification, the reduced sensitivity characteristic of hearing loss, and they generally incorporate mechanisms to protect against amplified intense sound. Recently developed digital hearing aids hold the promise of also providing more advanced forms of signal processing that will overcome reduced resolution for sound differences that restrict communication. For example, a number of promising techniques for improving communication in noisy environments are currently being evaluated for possible inclusion in digital hearing aids. Also, the possibility of developing an implantable hearing aid is being studied. Such a device might offer increased freedom from acoustic feedback and increased functional gain, as well as greater convenience and cosmetic acceptance. Achieving these goals could result in the effective use of hearing aids by a larger proportion of the impaired population. To fully exploit these advances, research on hearing aid selection procedures need to be expanded to ensure that each impaired listener is provided with the most appropriate device.

For individuals whose loss of auditory function is essentially complete, communication typically relies on the use of alternate senses. Recently, great progress has been made in the development of devices that stimulate the auditory system electrically in response to sound. A few individuals are able to use such electrical implants to achieve remarkably good acoustic reception of speech, and many others are able to use them effectively in conjunction with speechreading. Moreover, the likelihood that the effectiveness of this strategy can be substantially increased in the relatively near future is great. Enhanced specificity of stimulation, more informative speech coding, and improved ability to predict the outcome of this approach are current topics of intense study. Additional research will clarify the suitability of this approach for treating deafness in very young children.

The potential for using the tactile sense to substitute for hearing has also been studied extensively. Wearable tactile aids are currently being used in some schools for the deaf, primarily to provide an awareness of environmental sounds and to supplement speechreading. Recent research results indicate that this supplementation can be effective compared to that provided by electrical stimulation systems. The potential to improve the effectiveness of these devices through increases in the number of stimulated sites and improved speech coding is great.

C. *Behavioral Approaches to Rehabilitative Management*

Speechreading is an important aspect of speech perception by hearing impaired people of all ages. Studies are needed of the visual and brain processes involved as

well as of the nature of the task and the mechanisms essential to successful speechreading. Evaluation of performance in the recognition and perception of speech is important both as baseline information, for purposes of diagnoses and planning for treatment, and as a measure of efficacy of training. New and improved methods of evaluating performance are needed, as are studies of the validity and reliability of tests currently used.

For adults, rehabilitation often consists of communication therapy, which includes counseling in assertiveness training and development of strategies to enhance communication with others. The procedures used in these programs should be evaluated with regard to their efficacy relative to other activities, such as speechreading, training in the use of telecommunication aids, and the use of such assistive devices as captioning and auditory training.

For children, the impact of hearing loss is often influenced by the ability of family members to participate in management of the hearing impairment. For this reason, family-oriented intervention is an essential component of the intervention programs for young children. Little is known about which components of this intervention program are the most effective in preventing psychosocial problems associated with hearing impairment and ameliorating other consequences of hearing loss.

Early intervention is widely recognized as important to the successful development of hearing impaired children, but it is not currently available to most families. The average age of identification of deaf children is 2 1/2 years. Because eventual competence in language is largely determined by age 3, top priority should be given to the development of programs that will detect hearing loss in the first few months of life and provide appropriate intervention on an ongoing basis.

Deaf Children

Children who are born with profound hearing impairments constitute a special population whose habilitative needs differ in many ways from those of children who have less severe hearing losses. The major difference between the two populations lies in the degree to which they can use auditory information to learn their native language.

The research needs associated with deafness center around the effects of deafness on all aspects of development. Auditory speech perception is extremely limited, language is severely delayed, speech is often unintelligible, psychosocial development is adversely affected, and educational achievement is extremely poor. In spite of more than 200 years of special education, the efficacy of the procedures used with deaf children and the manner in which these procedures interact with their individual learning styles and cognitive status is

largely unknown. Clinical trials in which different teaching strategies, settings, and materials are evaluated have not been conducted. Such trials should constitute a major area of research over the next decade. Included in such studies should be the efficacy of the use of cochlear implants as well as the use of other sensory aids.

Deaf children of deaf parents constitute a special subgroup of this population. Because such persons are exposed to signed language from birth, they do not suffer the same type of language deprivation as do other deaf children. They often develop signed language skills at the same rate as normally hearing children. However, their learning of English is delayed. Indeed, they face a task much like that of learning a foreign language without access to sound as a learning tool. Therefore, there needs to be an effort to investigate the ways in which a signed language, such as American Sign Language, can be used to facilitate the development of the ability to use English, especially in its written form. A long-term, intensive study of ways to enhance the reading skills of deaf children (both of deaf and of hearing parents) should be supported.

Hard-of-Hearing Children

In contrast to deaf children, hard-of-hearing children have usable hearing that can be aided significantly by hearing aids. Such children are often educated in regular classrooms with and without special assistance. Very little research has been conducted with this population of children.

Some major areas of research to be accomplished in the next few years are:

- Determine effects of early amplification and intervention on development of speech, language, educational, and psychosocial skills.
- Determine effects of different educational settings on performance.
- Evaluate efficacy of therapy programs currently in use (auditory training, speech training).
- Develop ways in which to minimize social problems associated with moderate hearing loss.

Identified priorities in this area are:

- Early identification and intervention with hearing impaired children, including clinical trials of the effectiveness of current educational and rehabilitative strategies.
- Sensory aid research designed to improve the devices used and to evaluate their effectiveness with hearing impaired individuals.
- The development of English language skills by deaf and hard-of-hearing children.
- Development of outcome predictions for users of cochlear implants, and evaluation of the efficacy of implants as aids to communication.

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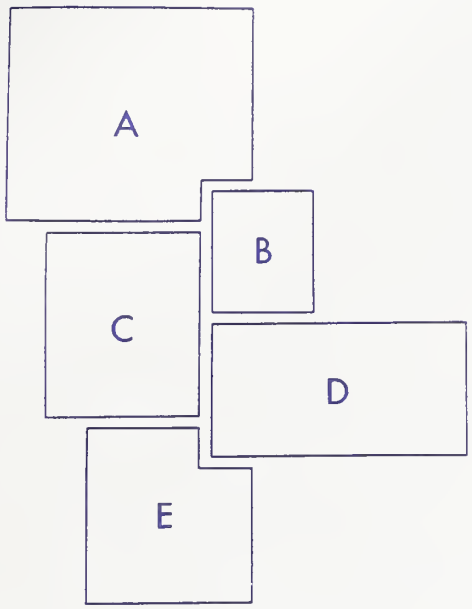

Cover:

A. Hearing impaired children, Boys Town.
National Institute for Communication Disorders in Children, Boys Town, Omaha, Nebraska.

B. The vitality of future research depends on the recruitment of research trainees.
National Institute of Neurological Disorders and Stroke, NIH.

C. Research on new ways to diagnose dizziness. Electrodes record eye movements while the balance system in the inner ear is stimulated.
National Institute of Neurological Disorders and Stroke, NIH.

D. A youngster improves the quality of his speech by watching and imitating his speech therapist.
Bill Branson, NIH.

E. Rotating, stimulus-sampling table to which methyl ethyl ketone odorant concentrations are fed from a 10-stage air-dilution olfactometer.
Richard L. Doty, Ph.D., Smell and Taste Research Center, University of Pennsylvania.